Skull Base Surgery: Strategies

Walter C. Jean

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Skull Base Surgery: Strategies

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To my mother

Dora Chu Jean
1941-1996

a lifetime dedicated to the education of her son

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Foreword

I believe this book serves to highlight a watershed moment in the history of skull base surgery and marks the exact moment in time when the primary practitioners of this art came together to describe a more conservative philosophy in the application of "the implements of war," which we call skull base surgical approaches. Perhaps the book is even a quiet apology to, or at least a nod in the direction of, all those patients whom we may have harmed unwittingly in our quest for innovation and perfection. The early description of surgical approaches to skull base tumors was met with unbridled enthusiasm by many young and eager neurosurgeons, myself amongst them, believing our generation to be the one to "beat the meningioma" and meet the challenge laid before us by Harvey Cushing. But surgical cures can be elusive even when dealing with benign tumors that have a grasp on neurovascular structures and take hostage the quality, if not the quantity, of a patient's life.

Our enthusiasm for these operations waned not through thoughtful introspection but through collision with undeniable barriers placed in our path. These barriers included outcome analysis, quality of life measures, and natural history, the mortal enemy of many a bold and daring operative conquest. However, the greatest blow of all to our naively overzealous pursuit is perhaps the patients' participation in determining the quality of their own outcome and survival. Until these barriers came to light, patients had surgery because the surgeon felt it was necessary, and their outcomes were acceptable because the surgeon said it was. At the same time these techniques were being developed, there was also another technique developing on a parallel path: radiosurgery. The skull base surgeons attacked radiosurgery and radiosurgeons with a vengeance for their "cowardly approach" to tumors while the rest of us were engaged in heroic, sometimes epic battles. One such battle in my early years went on for twenty-eight hours, through three changes of shift, and in the last few hours felt like a battle being fought in quicksand. Somehow the patient survived, through no credit of my own. Eventually, surgeons relented and adopted radiosurgery, not as a rival, but as an ally against both the onslaught of the disease and the shortcomings of our surgery.

This is not a call to pull back on the development of our skills, our innovation, or our courage going into battle. When we enter the operating room, as we often must, we must be sharp, we must be determined, and we must be courageous. But it is an admonition to be sure we always place the interests of our patients above our own—above our interest to prove our surgical prowess, to prove our point of view, to prove our inventiveness. Remember the phrase, "Physician Know Thyself."

I know that at some point in a difficult tumor surgery, if the battle between me and the tumor becomes personal, the patient becomes an innocent bystander to events. Sometimes I write a message to myself on the operating room board to remind me who it is we are operating on and what they want: "This is a single mother of two children. Without her, they are orphans;" to serve as a reminder, in case I lose my way, that this is not about me.

I think that it must be impossibly hard sometimes for soldiers under fire to adhere to the "rules of engagement" and make profound sacrifices to protect the innocent. When surgeons enter the operating room, we, too, go into battle, and we, too, must be prepared to make profound sacrifices to protect the innocent, our patient: we must sacrifice our ego and resist the siren's call of fame.

Harry R. van Loveren, MD

Preface

"How do we get there?" I have been asked this question innumerable times by students, residents, and colleagues, as they ponder an image of a skull base tumor on a radiographic study. The central idea of this book is to take you through the decision-making process of choosing and executing a surgical approach for skull base tumors: from the assessment of the clinical presentation, to the appreciation of the nuances of the anatomical details on the diagnostic images, to the analytical thinking process of designing the operation. With variations in size and shape, differences in biological characteristics, unique anatomic locations and extensions, and particular relationships and entanglements with nerves and vessels, there are infinite ways skull base tumors are different from each other. Obviously, it is impossible to teach an infinite number of operations to remove these tumors. But, just as the English alphabet only has 26 letters and more words than one person can master, once we break down the operations into component parts, the process of designing an operation becomes teachable.

Which approach corridor to use, which craniotomy opening to make, which additional bony elements to remove… these are the building blocks for every skull base operation. Experienced surgeons mix and match these building blocks to design an operation, and they do so subconsciously. Teaching one how to perform each element is necessary but insufficient to train a skull base surgeon. It is equally important, if not more so, for surgeons to learn the analytical process of how to combine the building blocks to tailor-make the operation to fit a specific patient and the surgical goal. With that, the process of designing a skull base operation is de-mystified.

Many existing books on skull base surgery show you what neurosurgeons do; they are atlases with beautiful dissections, but limited, if any, discussion on the practical applications of the surgical steps. In others with more verbal content, the chapters are headed by disease entities, and the chapters flow from a discussion of the disease topic to generalized approaches for its surgical management. This book is very different. It focuses not only on what skull base surgeons do, but also equally on how they think and strategize. The chapters flow from clinical presentation and radiographic/anatomical findings of a specific patient and tumor to the decision-making process and execution of the surgical approach. Obviously, no military general would create a battle plan and then seek an enemy to fight with those plans. A strategist sees an enemy, analyzes its strength and weakness, investigates the intervening terrain, and then formulates a battle plan. Similarly, a surgeon meets a specific patient, analyzes the patient's clinical data, reflects upon his or her own surgical training and experience, and then decides what to do. The flow in each chapter of this book aims to follow this sequence, and the pedagogical process is achieved though real-life case examples, not through generalized or theoretical discussions.

Finally, it is important to highlight that strategic thinking and decision making in skull base surgery is not about finding the unique solution to the clinical problem. Unlike theoretical mathematics, in skull base surgery the path to the truth is seldom singular. There exist no two surgeons with the same training, similar successes, or identical failures. As such, each chapter of this book will have a concluding section with a different voice from another surgeon with a unique point of view. The aim is not to critique the ideas of the main chapter, nor to set up a debate on some controversy such as "endoscopic vs. open" approaches for a particular tumor. The Perspectives section is placed at the end of each chapter to embrace the variations in the "surgeon factors," to look at the same topic from a different angle.

Each chapter is intended to stand alone as a complete unit, to minimize the labor on your part referring back-and-forth between them. Moreover, different authors offer diverse perspectives on similar ideas, techniques, and approaches. I hope, therefore, that you will excuse some minor repetitions amongst certain chapters, and that these do not amount to redundancy.

Walter C. Jean, MD
Washington, DC, 2018

Acknowledgments

When the concept of this book was solidified, I was confronted with the daunting task of assembling a large cast of writers who are not only maestros in the art of skull base surgery, but who can also eloquently explain their thought process and meticulously describe their intricate technique to eager learners. My greatest fear was that few would be interested in participating, and thus even fewer would be predisposed to acquire the final product if it contained only my monotonous voice droning on chapter after chapter.

My fear dissipated when master-surgeons signed on, one by one. As the discourse in 21st century skull base surgery becomes increasingly global, I thought it important that the book contains ideas from people whose mother-tongue is not English, even though it is the base language of the publication. I am extraordinarily humbled by the colleagues from around the world, from Japan to Italy, from China to Brazil, who patiently recorded their hard-earned knowledge and insights in their second or third language and, additionally, by all the writers who graciously opened their vast archives of surgical travails and shared the most instructional cases with our readers. They also brought along a cavalcade of their prodigious disciples, whose diligent work fills the pages that follow. These learners-turned-teachers will assuredly become the next generation of skull base virtuosos and hopefully find good use for this book as their careers progress.

To have the privilege of steering this project, I am indebted first and foremost to my trainees, both former and current, for their challenging questions and constant curiosity. Working through complex clinical problems with them, and justifying my own conclusions, is the best way to continue my own learning, which will no doubt be a life-long process. My various colleagues and previous mentors deserve obvious gratitude, as without their belief in me I would have no career, let alone the chance to lead a project such as this. However, one of my teachers deserves the spotlight. Many of the contributors of the following chapters, including Professors Aziz, Youssef, Huang, Agazzi, van Gompel, Froelich, and Link, have also learned from the unparalleled skill and articulate lessons of the legendary teacher Harry van Loveren. As a master strategist, his instructions continue to permeate my decisions on a daily basis, whether in the operating suite, clinic, classroom, or in front of the computer.

I am thankful to my chairman, Dr. Anthony Caputy, and my colleagues at the George Washington University, for their support initiating this project and throughout the entire process. Similarly, I owe tremendous gratitude to my wife and two boys for indulging daddy working on his laptop from ski lodge to tropical resorts, sending emails during Santa's visit as well as weekends when our only callers were birds in the backyard, and even revising a figure with the production team, on the phone, on a ride, at Disney (yes, this actually happened).

My gratitude goes to the talented illustrator Jennifer Pryll, whose virtuosic artistry is surpassed only by her ability to decipher my arcane anatomical descriptions and transform my squiggles into works of art, even the ones shared with her from "It's a Small World." Finally, I must thank the staff at Thieme, and especially Timothy Hiscock and Sarah Landis, for the critical roles they played in transforming the writers' ideas into the book that is in your hand.

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Introduction: Modern Skull Base Surgery and Timeless Strategic Philosophy

Walter C. Jean

■ How Do We Start?

The Goal: *Three* Factors

Sun Tze was a Chinese general and philosopher, active during the tumultuous late Chou dynasty around 500 B.C. His masterpiece, "bin' fa," which loosely translates to "soldiering way or method," is widely known in the Western world as "Art of War." He wrote,

> *...the general who wins a battle makes many calculations in his temple ere the battle is fought...do many calculations lead to victory, and few calculations to defeat... It is by attention to this point that I can foresee who is likely to win or lose.*

The most important "calculation" that one must make before designing an operation is the enunciation of the over-arching surgical goal. Just as the overall aim of a war will influence how individual battles are fought, the surgical goal will affect the design of an operation. As such, the main reason to set the surgical goal is so that one can tailor-make the operation to achieve those specific goals. An example of how the goal dictates the approach is illustrated by the patient whose MRI is shown in. **Fig. I.1.** This was a 74-year-old woman with active breast cancer, presenting with worsening balance. Surgery was necessary to halt her neurological deterioration, and while her co-morbidities limited the surgical goal, they do not nullify them. But, whereas a complex petrosectomy might be the right approach for a radical resection, a subtemporal/transtentorial approach is perhaps the better choice for her given the more rational goal of partial resection for brainstem decompression.

> *...just as water retains no constant shape, so in warfare there are no constant conditions...*

Since no battle plan ever survives an encounter with the enemy, the fluidity of the operation is another important reason for setting a firm goal. As an operation progresses, predictable changes in the anatomy during the approach and tumor removal may lead to unpredictable events that distort the calculations previously made. Events such as the disruption of an important blood vessel, fluctuations in the monitored brainstem potentials, or the discovery that the tumor is hopelessly adherent to a cranial nerve can alter what the surgeon does, but they do not change the aim of the operation. At such moments when pre-made plans are disrupted by events or findings during the operation, it is critically important to focus on the goal of surgery when adjusting those plans. If the goal has yet to be reached, then one must proceed and find alternate ways to achieve the goal. On the other hand, if, for example, the goal of surgery was "brainstem decompression," and that has already been achieved, then continuing the operation for a complete tumor removal under those conditions may risk further or more serious complications.

The variables that enter the calculations for the surgical goal generally fall into three types: patient, anatomic, and surgeon factors. Patient factors are relatively easy to understand, and most surgeons learn to do these calculations in medical school. Generally speaking, these factors include conditions and occurrences in the patient's past, such as cardiopulmonary co-morbidities, and the treatment history of the patient's tumor (**Box 1**).

> *In the practical art of war, the best thing of all is to take the enemy's country whole and intact; to shatter and destroy it is not so good.*

The achievement of a cure is a holy grail for physicians, but as modern medicine can still only manage such common diseases

Box 1. Patient Factors

- Pre-existing health issues such as cardiovascular or pulmonary disease
- Use of anti-coagulation
- Previous treatment for the same tumor such as prior surgery or radiation therapy
- Psycho-socio-economic factors that may impact on recovery period

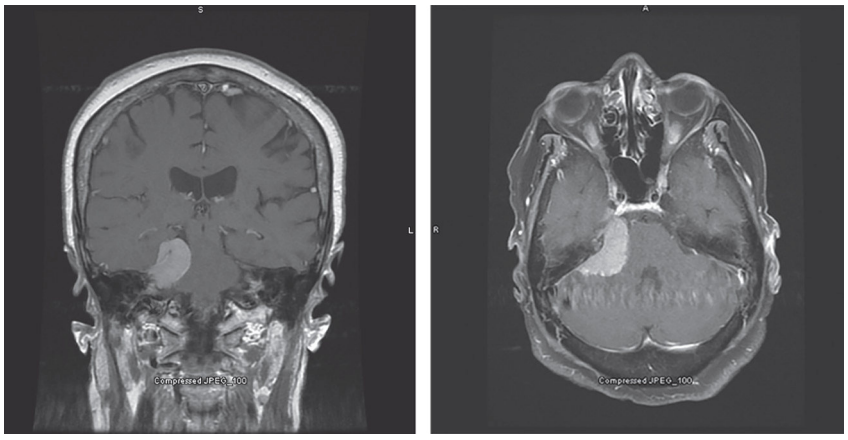


Fig. I.1 MRI of a 74-year-old female breast cancer patient with deteriorating balance.

like diabetes, a cure remains elusive for most problems. Skull base surgeons chase similar holy grails of complete resections with pristine postoperative MRI scans, but none of that is meaningful if the patient is harmed. Careful consideration of the patient's age, co-morbidities, and treatment history will prevent a well-intended operation from hurting the patient.

Box 2. Anatomic Factors

- Size, location, and histopathological identity
- Does the tumor engulf critical vessels or nerves where it should be left alone?
- Does it exist in an area where it should not be entered (e.g. cavernous sinus with healthy cranial nerves and carotid artery)?

Water shapes its course according to the nature of the ground over which it flows; the soldier works out his victory in relation to the foe whom he is facing.

Working out this relation with the "foe" requires understanding the intricacies of anatomy. Anatomic factors are often subtler than patient factors, and surgeons spend their years in training learning to make these calculations, such as considering the size and location of the tumor, where it is protected from approach by the bony skull, and where it might be more exposed for approach (**Box 2**).

There are roads which must not be followed…towns which must not be besieged, positions which must not be contested…the power of shrewdly calculating difficulties, dangers and distances, constitutes the test of a great general.

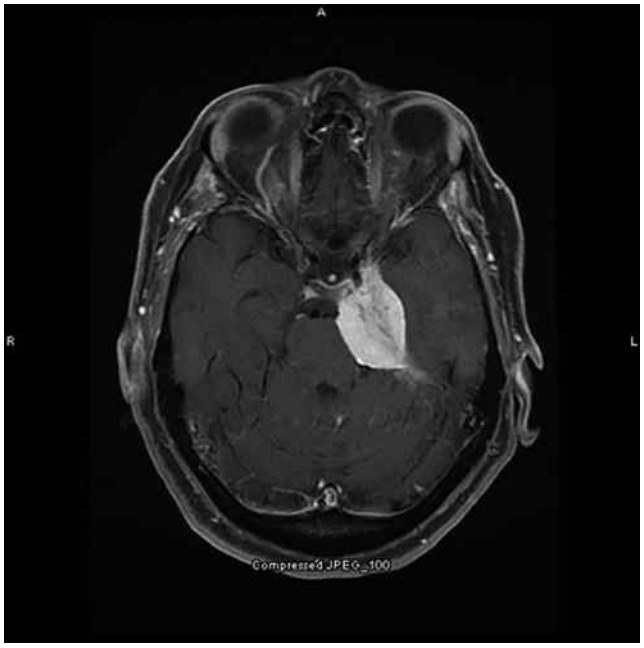


Fig. I.2 MRI of a cavernous meningioma with radiographic progression over several years.

Tumor location is perhaps the single most important anatomic consideration for setting the surgical goal. For example, the MRI in **Fig. I.2** belonged to a 64-year-old woman with asymptomatic, radiographic progression of her tumor (**Fig. I.2**). The tumor compressed the pons and involved the left cavernous sinus. Whereas brainstem decompression is rationally necessary, risking oculomotor dysfunction and carotid injury by removing the cavernous portion is unwise in this asymptomatic woman nearing her 70s.

Consideration of the tumor type is another important aspect of the anatomic calculations. A poignant example of this is shown in **Fig. I.3**. The majority of adult cerebellar tumors are metastatic in nature, and these are most commonly removed by entering to debulk the mass and resecting the tumor piecemeal from center outward. However, the most common primary tumor in this area is a hemangioblastoma, and these tumors must not be entered and cannot be debulked without massive blood loss. Instead, one must work around the periphery and resect from the outside moving inward to remove the tumor intact. If a diagnostic angiogram confirms the suspicion of such a hemangioblastoma, then the goal must be set for such an en bloc resection.

If you know the enemy and know yourself, there will be one hundred victories in one hundred battles

The third set of factors is related to the surgeon, and these are the most difficult to calculate. It requires honesty and significant self-awareness, which some of us acquire only late in our careers. There are questions that we must ask ourselves before every operation, such as: "Do I have enough experience and training to do what is necessary?" or "Should that last devastating complication stop me from doing this particular operation again?" There are no books, mentors, or outside resources that can make these calculations, for these are immensely personal, depending on what we have learned in the past (**Box 3**).

Box 3. Surgeon Factors

- Training and experience to do what is necessary
- Personal experience with complications
- If a multi-disciplinary skull base team is needed, do I have this in place?
- Skill and experience of other team members in the operating room, such as anesthesiology, nursing, and monitoring staff

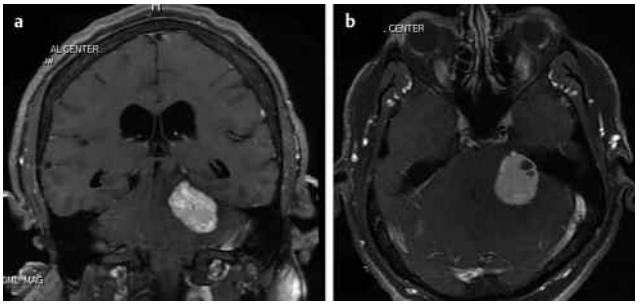


Fig. I.3 An adult cerebellar tumor. (a) Coronal and **(b)** axial MRI with gadolinium showing a large enhancing lesion. Even though the cystic portion is small, the tumor was a hemangioblastoma.

Needless to say, surgical training can vary widely between surgeons, but we also experience failure at different times and in different ways. Successes in training and failures in complications are the yin/yang basis of our learning, but the latter may be more significant. First of all, they are unique. No two surgeons can possibly experience the same complications, and thus our failures are personal. Secondly, whereas we are able to suppress our emotions almost by necessity to survive the hardship of residency training, our complications pack much heavier psychological punches. Rene Leriche's oft-used quote comes to mind here: "Every surgeon has a cemetery where he goes to pray…to look for reasons for his failures," noting that our individual way of interpreting failure adds yet another layer of divergence in what we learn. We must decide individually how bad of an outcome from a particular operation or how many complications from a certain approach should give us pause for repeating it again. So, while failure is universal, learning is unique, and no two surgeons can ever consider the same problem from exactly the same perspective.

■ How Do We Get There?

The Approach: *Three* Elements

Military tactics are like unto water; for water in its natural course runs away from high places and hastens downwards. So in war, the way is to avoid what is strong and to strike at what is weak.

For a deployment of soldiers, a flank unprotected by fortification or natural barrier is often described as "up in the air" and vulnerable to assault. Similarly, a tumor may have an exposed side, unprotected from critical fiber tracks or eloquent cortex, where it might be best attacked. Cystic portions of a tumor near a cortical surface is one example of such a weakness, as approaching the tumor through the cyst often provides significant freedom for maneuvering instruments and working space for the operation.

Size, configuration, location, histopathological identity, relationship with vessels and nerves…different combinations of all these characteristics determine where the tumor is best attacked. Subtle variations on any of these features alone can also impact the choice of approach in major ways. Logically, to best eliminate these tumors, we need infinite variations in surgery as well. But how do we learn infinite numbers of operations? Would that not overwhelm the learner and exhaust the teacher? The key is to understand that, just as a combination of five characteristics account for the diversity in the tumor, it is a similar combination of surgical approach elements that allows us to tailor the perfect operation for each.

There are no more than five cardinal tastes (sour, acrid, salt, sweet, bitter), yet combinations of them yield more flavors than can ever be tasted. In battle there are not more than two methods of attack-the direct and indirect; yet these two in combination give rise to an endless series of maneuvers.

The "direct" method is simply a frontal assault directly into the enemy. For brain tumors, one sees this direct trajectory simply by drawing the shortest, straightest line from the center of the tumor, through the least amount of brain tissue, to the skull surface. Simple in concept, this direct line of attack is applicable for convexity tumors, but for most skull base tumors this may blaze a trail of destruction through critical neurovascular structures, making it completely unacceptable.

Thinking Inside-out: Corridor, Craniotomy, Modifiers

It is intuitive to design the surgical approach outside-inward, since that is how surgery is conducted, from incising the skin, to drilling the bone, to intracranial dissection. However, for the design of the operation, it is often easier to think the other way around, from inside-outward, by asking the hypothetical question, "What is the easiest way for the tumor to slip out of the head?" Following this method, the first consideration of the "indirect attack" for creating the conduit of the tumor's "exit" is the surgical corridor.

The natural formation of the country is the solder's best ally.

Surgical corridors are the "natural formation" of anatomical planes in the brain itself or within the skull/brain interface, which can lead the tumor to its exit. Whereas the direct route is the shortest, straightest line from the tumor to the skull surface, the corridor can be considered the "indirect" route, less destructive and subtler. As most of these corridors involve the interface between brain and skull, not surprisingly, many of them follow dural surfaces, whether they are adherent to bone or more centrally-located structures such as the tentorium or falx cerebri.

Sometimes, these indirect routes are quite obscure. Consider the tumor shown in **Fig. I.4**. At first glance, the tumor seems

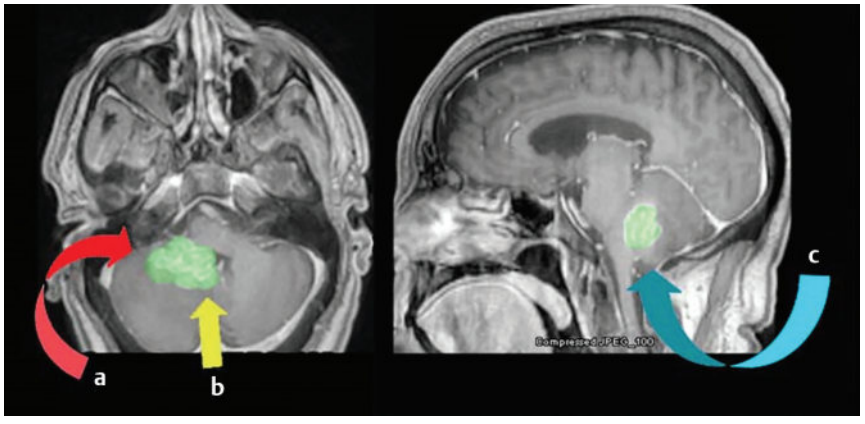


Fig. I.4 Options for surgical approach for tumor resection. Conventional approach options include **(a)** a lateral-to-medial trajectory, which would pose challenges to reach the midline portion of the tumor, or **(b)** a medial-to-lateral trajectory which would entail reaching the tumor by going through a significant amount of normal tissue. The inferior-to-superior approach through the subtonsillar corridor shown in **(c)** was chosen because it offers the best exposure of the tumor with the least tissue destruction.

to be protected on all sides by the cerebellum. Approaching it from a medial direction would require damaging healthy tissue, and coming from a lateral direction might put cranial nerves in harm's way. But when one notices that the tumor is "vulnerable" in the cerebellomedullary fissure, exposing it through this natural anatomical cleavage plan seems completely logical, as it is the most direct route to the tumor without traversing healthy tissue.

The most commonly used approach corridors for skull base microsurgery are listed here (**Box 4**). As there is a finite set of these corridors, conceptualizing a complex operation just became much easier: to begin the design of the optimal approach, you start by selecting the shortest, straightest corridor that can be utilized for a particular tumor. This is the first tier of "approach element" that we will combine with others to design each operation.

Box 4. Common Approach Corridors

- Anterior Corridors
  - Subfrontal
  - Interhemispheric
- Anterolateral Corridors
  - Lateral subfrontal
  - Transsylvian
  - Transcavernous
- Lateral Corridors
  - Subtemporal
  - Transtemporal
- Posterolateral Corridors
  - Presigmoid
  - Transigmoid
  - Retrosigmoid
- Posterior Corridors (supratentorial)
  - Occipital interhemispheric
- Posterior Corridors (infratentorial)
  - Infratentorial/supracerebellar
  - Transvermian
  - Subtonsillar
- Transventricular Corridors
  - Transcortical
  - Transcallosal
  - Precuneus

From the site of the tumor, the surgical corridor leading outward eventually arrives at the skull. For different groups of corridors, there exists a basic bony opening, or craniotomy, that serves as the corridor's standard "front door." For example, the standard doorway to the anterolateral corridor, which is used commonly for approaching the juxtacavernous or sphenoid wing region, is the pterional craniotomy; whereas for a posterolateral corridor, the standard opening is the retrosigmoid craniotomy. These doorways or openings are the second tier of approach elements we need to consider, but as any particular corridor almost always lead to the same front door, selecting the proper craniotomy to open any chosen corridor is usually quite simple (**Fig. I.5**).

A far more complex decision comes next. Some doors are locked, and you may need a key or passcode; if the door is guarded, you might even need a passport. The standard "bone opening" to a surgical corridor may be insufficient, and it is the embellishments or modifiers to these basic openings, in their many permutations and combinations, that afford the surgeon flexibility in the approach to tailor-make the perfect operation. These third-tier approach elements mostly involve additional bone removal to widen the standard doorway to the corridors. For the standard pterional craniotomy, these modifiers may include removal of orbital structures, the zygomatic arch, or the anterior clinoid. For the middle fossa craniotomy, these embellishments may include maneuvers such as the anterior petrosectomy or incision of the tentorium.

If you consider these three tiers of approach elements as building blocks, like cardinal tastes, musical notes, or primary colors, which can be combined in infinite variation, you can conceptualize the design of the perfect approach without becoming overwhelmed by choices.

Box 5. A Demonstration of the Method

In this book, various master surgeons will discuss the thought process behind how they design operations. As such, I will only briefly demonstrate the application of this method with one example. **Fig. I.6** shows a petroclival meningioma in a young man with progressive pain and a debilitating balance problem. The goal was set for brainstem decompression, with a secondary goal of removing as much of the tumor as possible. The straightest, shortest route to tumor exit appeared to be the posterolateral **corridor,** with the associated "standard" **craniotomy** of a retrosigmoid craniotomy. The tumor involved the clivus and reached past the midline on the left. It extended from a level just above the internal acoustic canal all the way to the foramen magnum. Because of this, the corridor and bone opening needed to be expanded by a posterior petrosectomy and removal of the occipital condyle. These are the **modifiers**. Putting all those "building blocks" together, one arrives at a "Posterior petrosectomy and transcondylar approach for combined pre-and retro-sigmoid resection of a petroclival meningioma" as one solution to the clinical problem with which this young man presented.

■ What Do We Do When We Arrive?

The Resection: *Three* Conditions
Controlling the Space

The good fighters … first put themselves beyond the possibility of defeat, and then wait for an opportunity of defeating the enemy.

Once sufficient exposure has been obtained at the end of the approach, controlling the surgical environment is the first task for the resection. This control allows you to put yourself "beyond the possibility of defeat," and it mainly has to do with protecting healthy neurovascular structures. Frequently, this task is as important if not more so than removing the tumor itself. It is certainly critical in avoiding devastating complications. Whatever hinders you from "controlling the environment," that blocks you from safeguarding critical structures, should be eliminated if possible. One such example is the

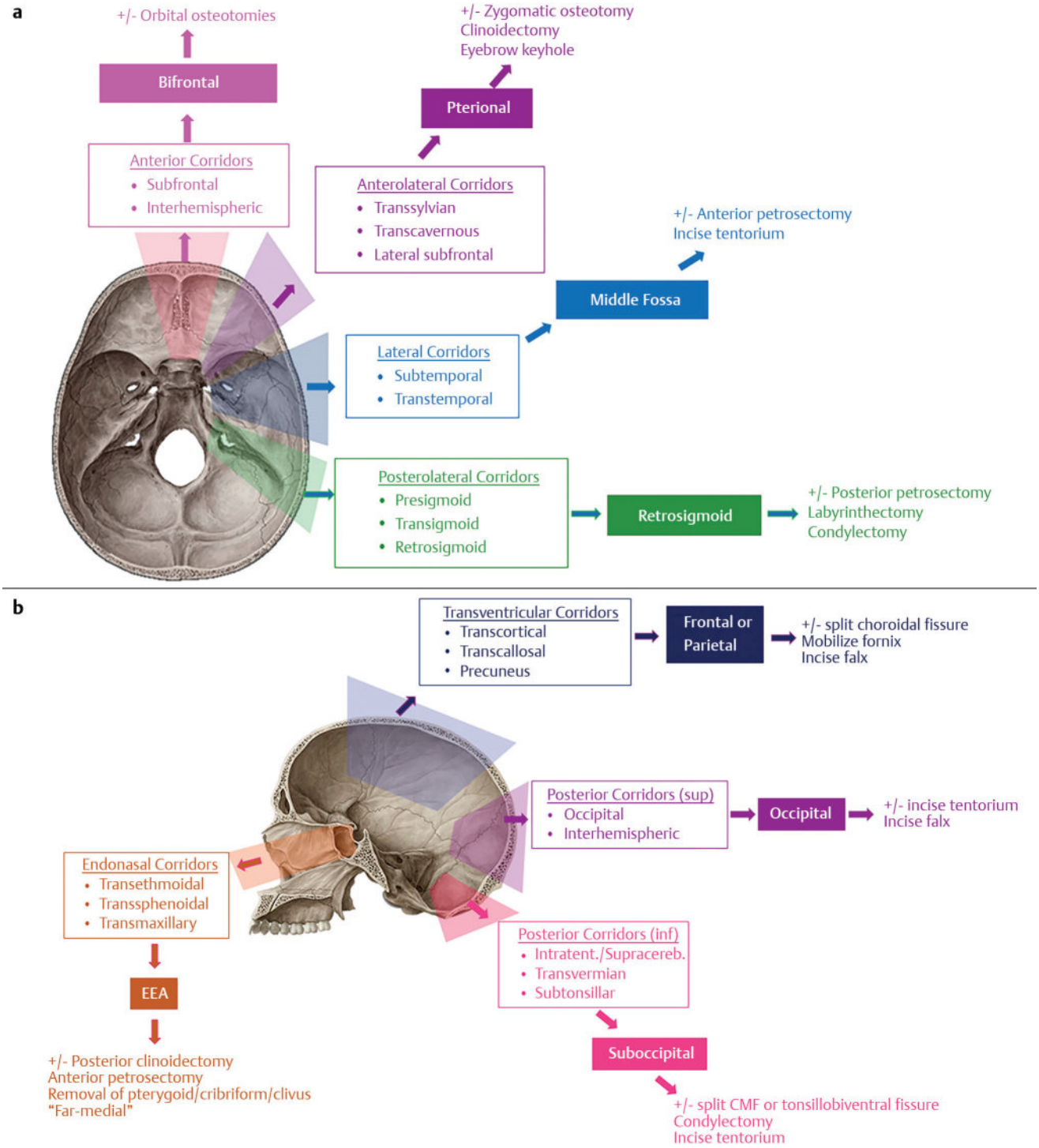


Fig. I.5 Skull base corridors and the three tiers of approach elements. (a) Elements in the axial plane and **(b)** in the coronal plane. Moving away from the region of interest, the first box contains the commonly-used surgical corridors to approach that region. Most of these corridors are natural anatomical cleavage planes, of which surgeons can take advantage to minimize tissue damage during surgery. Moving away from the first box, the next element is the standard bone opening utilized to access those corridors. Moving further away, the third and last elements are "modifiers" of the bone opening that can expand or otherwise improve the opening to the corridors. All the elements should be used in different combinations to design skull base operations. Notice that even though surgery proceeds from the outside in, the decision-making process for designing the operation works inside out, with the arrows starting near the target pointing outward.

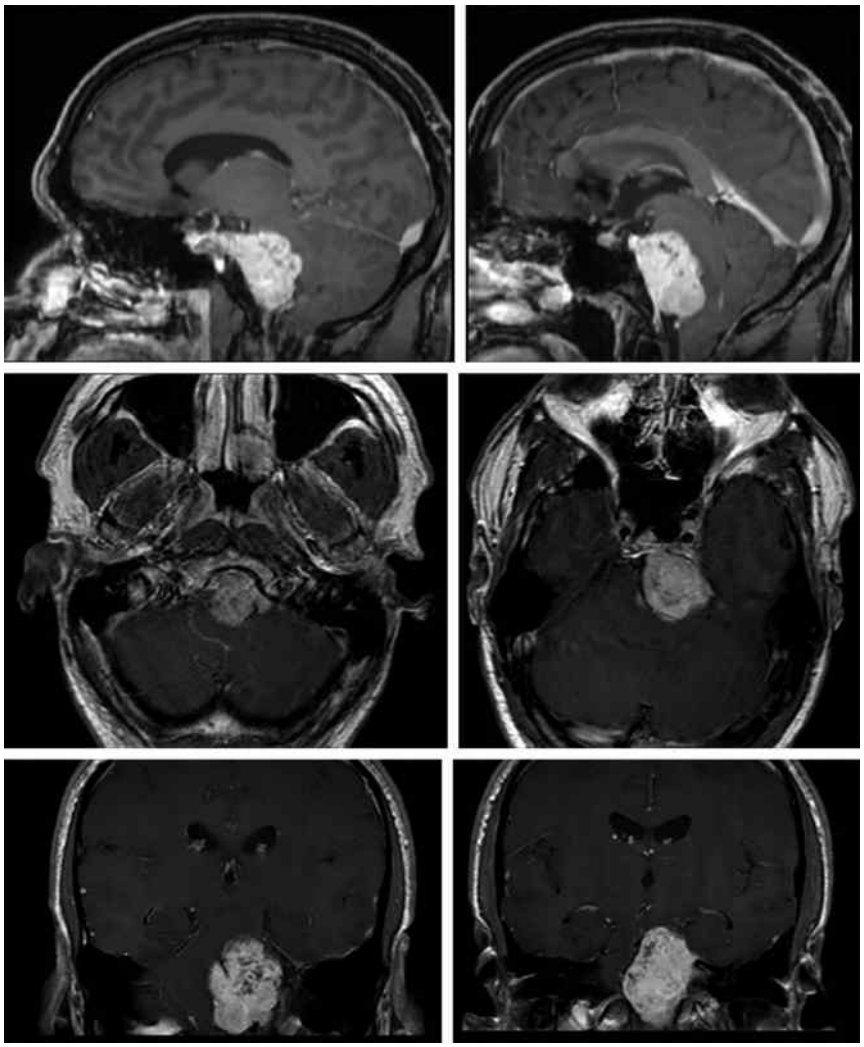


Fig. I.6 MRI of a young man with headache and debilitating balance problems: demonstration of the technique using corridor, craniotomy, modifier.

tentorium or falx, which can be safely incised to gain more freedom to maneuver.

If precious structures can be protected right away, then that should be done next. For example, during an anterolateral skull base approach to a craniopharyngioma, the ipsilateral carotid artery and optic nerve may be seen very early, before the tumor resection. They should be disentangled from the tumor and safeguarded immediately for control. Sometimes, this protection occurs later. An example of this is the facial nerve during a translabyrinthine approach to an acoustic neuroma. If the tumor is sizable, then tumor debulking must take place first, and this allows for identification of the facial nerve at the brainstem, which can then be protected.

Often complete control is not possible, and if a critical and vulnerable structure cannot be protected directly, avoiding "defeat" involves constantly reminding yourself to stay away from it as the tumor is removed. An example of this is the carotid artery "hidden" in the cavernous sinus during resection of a juxtacavernous tumor. "Stay out of the cavernous sinus," like a Buddhist chant, should be droning in the background of your mind as the resection proceeds.

In general, each surgical move should make the next move safer. An example of this is the handling of feeding vessels. Consider the tumor in **Fig. I.7**. Endovascular treatment was

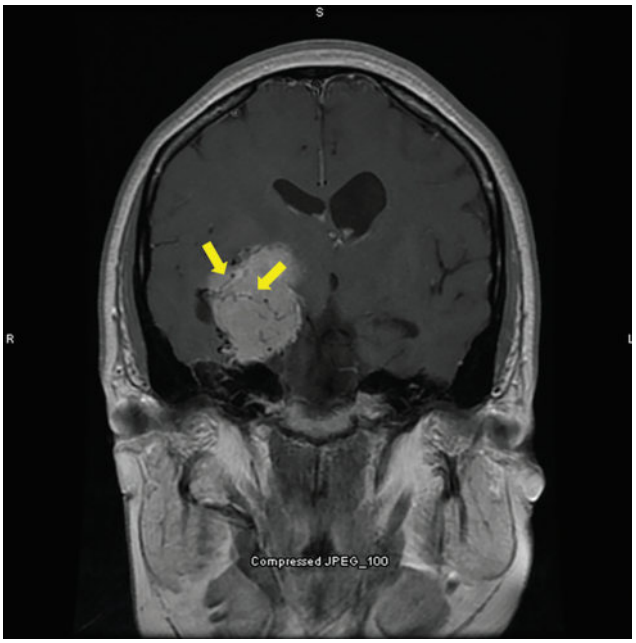


Fig. I.7 MRI of a large middle fossa tumor. Yellow arrows: large tumor-feeding vessels emanating from the choroidal blood supply.

not able to eliminate all the arterial supply to this tumor, and some of this supply reached the tumor on the ventricular surface. If, before the resection begins, a transventricular access is performed to eliminate these feeding arteries, it would make the resection move along much more efficiently with less blood loss. This example also raises the point that every complex operation for a skull base tumor always turns into a vascular operation at certain junctures. Readying temporary aneurysms clips for use is rarely a bad idea.

If the tumor's blood supply is significantly reduced, debulking the mass may seem far more mundane than the preceding steps, but like other parts of the operation, this must proceed with a plan. As the tumor mass is removed, the brain may "reclaim" some of its space, and the surgical window progressively narrows. Debulking must therefore proceed in such an order to avoid premature closure of the working space. Frequently, that means the deepest portion of the tumor must be removed first, as the rest of the tumor bulk acts as a "natural retractor" to stent open the working corridor. As removal of the mass continues, ideally, the tumor "delivers itself" into the space that is vacated, and the resection can proceed through progressively smaller windows. Conversely, the temptation to widen the operative window by deepening the retractors should be avoided.

Managing the Time

... though we have heard of stupid haste in war, cleverness has never been seen associated with long delays ... In war, then, let your great object be victory, not lengthy campaigns.

A surgical move that makes the next move safer also makes it more efficient. As the example in **Fig. I.7** already highlighted, elimination of the arterial supply automatically makes the resection move faster as it mitigates bleeding (**Fig. I.7**). Another aspect of speed is even more important to understand. Achieving speed in surgery does not require doing anything quickly (i.e. "stupid haste"), but instead requires efficient transition between moves without wasting time. This efficiency, in turn, requires anticipation of transitions. Having a clear operational plan focused on accomplishing the surgical goal allows for this anticipation.

Although every part of an operation is equally important, there is a logical pacing of each segment of the procedure such that some move faster than others. It is important to follow this natural flow. During the tumor resection, for example, the debulking steps should move expeditiously, and the efficient completion of these steps preserves the energy and concentration of the surgeon. This reserve can then be spent when it is necessary during the slow down at the "end game," when the last parts of the remaining tumor are dissected away from critical structures such as the brainstem or cranial nerves.

... the opportunity to defeat the enemy is provided by the enemy himself ...

Speed is also achieved by recognizing the precise moments to move aggressively. There comes a time, almost inevitably in every operation, where the tumor yields an opportunity for its own "defeat," and it is critical for the surgeon to seize this. The classic example is when, after seemingly endless minutes of debulking the mass, a plane of separation appears next to the tumor capsule. If squandered, this opportunity may be lost for the rest of the operation as surgical debris, such as clotted blood, obscures this plane. The seasoned surgeon seizes the momentum that the tumor yields, marks and preserves the separation plane to his or her advantage as the resection moves on.

Tempering the Zeal

... in war the victorious strategist only seeks battle after the victory has been won, whereas he who is destined to defeat first fights and afterwards looks for victory. The supreme art of war is to subdue the enemy without fighting.

Whether the surgical goal is excisional biopsy, decompression of the brain, separation of a target from a cranial nerve in preparation of radiosurgery, or complete resection, serious consideration should be given to halt the resection once the goal is reached. With an approach well-designed and a goal rationally-set, the surgeon should have little strife to finish the operation. In other words, if surgery is well-tailored, there should be no struggle, or "fighting," before the goal is achieved. Pushing beyond that goal either means that the goal was not set properly, or the surgeon is beginning to fight. Beware that allowing the operation to degenerate into combat between you and the tumor can lead to catastrophe for the patient. Achieving the designated goal is for the patient; fighting beyond that is for the glory of the surgeon.

... to hear noise of thunder is no sign of a quick ear. What the ancients called a clever fighter, is one who not only wins, but excels in winning with ease. Hence his victories bring him neither reputation for wisdom or credit for courage. He wins his battles by making no mistakes.

Fighting beyond the goal may not be the mistake itself, but mistakes happen in this process, and these often lead to devastating consequences for the patient. And having repeatedly highlighted the similarities of warfare and surgery, I must now emphasize the one major difference. In the former scenario, passion of the combatants is critical for the determination of the victor, whereas in the latter, passion often leads to blurring of the goal. As surgeons, we must keep our own emotions out of the operation because ultimately, it is the patient who will suffer the consequences of our zeal.

Part I

Tumors of the Anterior Skull Base

1 Tuberculum Sellae *3*

2 Olfactory Groove *18*

3 Nasopharynx and Pterygopalatine Fossa *29*

1 Tuberculum Sellae

Gordon Mao, Alexander Yu, and Khaled M. Aziz

Keywords: meningioma, transpalpebral approach, orbitofrontal craniotomy, endoscopic endonasal transtuberculum approach

■ Case Presentation

A 70-year-old female Caucasian elementary school teacher presented with the chief complaint of vision problems in her right eye. For over a year, she has had gradual decline of the vision in that eye. She had initially seen an optometrist, who documented bitemporal visual field defects and right optic disc pallor, and sent her for a neurology consultation. After confirming the findings, the neurologist sent her for a magnetic resonance imaging (MRI) which showed a large, avidly enhancing 2.4 × 2.0 × 2.4 cm dural-based mass draping over the suprasellar cistern consistent with an anterior skull base meningioma (**Fig. 1.1a–c**).

Questions

1. Given the tumor's location, what neighboring neurovascular structures may be directly involved with or infiltrated by the lesion, and therefore susceptible to injury during surgery?
2. If surgery is contemplated for this lesion, what would be your goal of surgery—biopsy, debulking, or complete resection?

■ Diagnosis and Assessment

The differential diagnosis of a sellar/parasellar mass includes pituitary adenoma, large aneurysms, optic nerve glioma, metastases, sarcoid lesions, craniopharyngioma, teratoma, and germinoma. However, given the appearance of this patient's mass with homogenous enhancement, extra-axial location, and extensive attachment to the dura with a dural tail, the most likely diagnosis was an anterior skull base meningioma. One would expect a craniopharyngioma to have more cystic components, and a pituitary macroadenoma to be centered over the sella turcica despite a suprasellar extension.

This patient's meningioma originated in the planum sphenoidale and tuberculum sellae area. Careful assessment of the anatomy and function of the surrounding neurological structures must be made before surgical plans can be considered. Her visual field had already been carefully documented. Displacement or encasement of the internal carotid artery (ICA), anterior cerebral artery (ACA), as well as the optic nerves and chiasm, must be noted in detail for surgical planning. In addition to an MRI, a computed tomography angiography (CTA) would often yield valuable information regarding these anatomical details, especially for large tumors.

In the absence of associated brain edema, or other evidence of brain invasion, this tumor was most likely a low-grade lesion. In this otherwise healthy patient, a complete resection (i.e., Simpson grade I or II) may therefore be curative. As such, a surgical goal for her was set to be a complete resection of the lesion with preservation of the adjacent critical neurovascular structures, including the optic apparatus, pituitary, anterior branches of the circle of Willis, and the cranial nerve within the cavernous sinus. Her visual symptoms indicated that special attention must be paid toward protecting and preserving her optic apparatus, to avoid any exacerbation to her already limited vision.

■ Anatomical and Therapeutic Considerations

Before selecting the operative approach, the anatomical nuances that may affect this choice must be considered. This patient's tuberculum sellae meningioma had a wide dural base, but it was not particularly "tall," measuring just under 2 cm in the superior/inferior dimension. On the contrasted MRI study, one can see that the optic chiasm was displaced posteriorly (postfix), and the optic nerves laterally. The tumor was in close proximity to the supraclinoid ICA and displaced the ACA superiorly, but there was no evidence of any encasement. Since the MRI was sufficient to yield this information, a CTA was not obtained for this patient. It is our practice to do so selectively unless the tumor is larger than 3 cm in diameter.

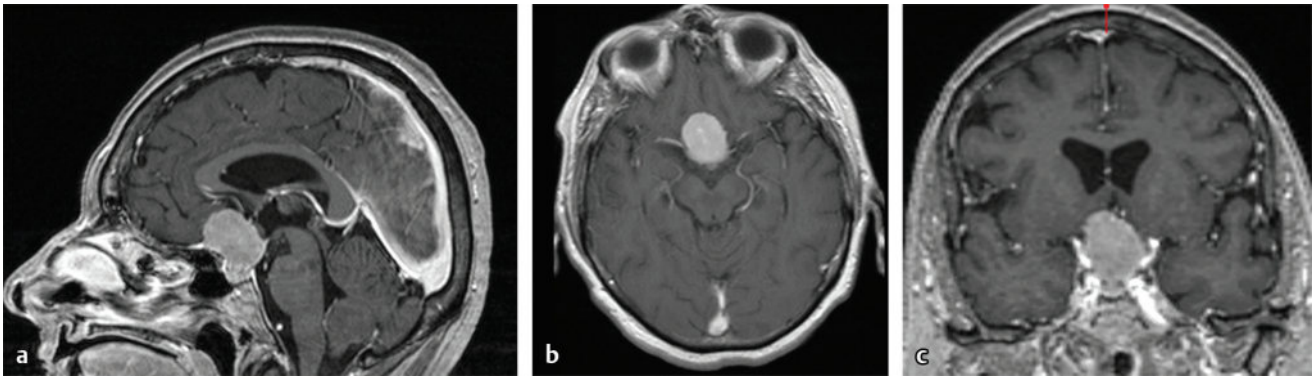


Fig. 1.1 Preoperative MRI brain postcontrast T1-weighted images. (a) Sagittal, **(b)** axial, and **(c)** coronal images showing the patient's tumor situated at the tuberculum sellae.

Options for Approach

The anterolateral corridor can be used to access this patient's tumor via a standard pterional craniotomy. This is the traditional "workhorse" approach used for lesions ranging from intracerebral aneurysms of both the anterior and posterior circulation, to pituitary macroadenoma and other common parasellar tumors. The main benefit of this approach is familiarity, since it is a standard operation practiced repeatedly during residency training regardless of subsequent subspecialization. The large craniotomy allows for excellent exposure of the entire parasellar region including the suprasellar cistern, opticocarotid cistern, and the dural base of the meningioma over the tuberculum sellae and planum sphenoidale. Direct access to the suprasellar cistern provides wide visualization of the infundibulum and optic apparatus and allows precise dissection and debulking of the mass without damage to these structures. An anterolateral approach also permits early identification of the ipsilateral ICA and ACA, reducing the risk of accidental injury of these vessels during dissection. Efficient elimination of the feeders from the anterior ethmoid artery during the approach can also potentially reduce the blood loss during the resection.

Another option is the endoscopic endonasal approach, used with increasing frequency in skull base surgery. With the latest optics and lighting in modern endoscopes, the parasellar area can be directly visualized and accessed without any brain retraction. In fact, the endonasal route may provide the shortest and straightest path to the tuberculum sellae target, and eliminates the need to work between nerves and arteries to resect the tumor. The tumor's blood supply is encountered first during the approach, and the elimination of this leads to a safer and quicker resection. The main problem of this approach for this patient's tuberculum sellae meningioma is in reaching the suprasellar areas, and injury to the pituitary in the sella can lead to postoperative endocrine dysfunction. Lateral extensions of the tumor abutting the cavernous sinus and carotid siphon make the dissection of the tumor from these sensitive vascular structures quite treacherous, and any injury to the carotids and branches would be very difficult to repair within such a small opening (**Fig. 1.2**). Perhaps with the exception for master surgeons, an optic foraminotomy from below to mobilize the optic nerve is very challenging. Tumor dissection without such a foraminotomy can easily stretch the optic nerves, leading to further damage to the patient's vision. Finally, we must consider the risk to the anterior cerebral arteries. Always draped on top of, and sometimes encased by the tumor, an endoscopic approach from below would reach these vessels only near the end of the operation. Since the ACAs are frequently obscured by the superior tumor capsule, they are prone to avulsion during the dissection of the final piece of the tumor, and again, repair would be extremely challenging.

Since the inception of minimally invasive approaches of skull base surgery in the past three decades, the supraorbital frontal craniotomy, with or without an orbitotomy, has evolved to become a versatile approach for all types of anterior fossa lesions. Sometimes called a "keyhole" approach, this variation of using the anterolateral corridor has the advantage of the most direct trajectory to the anterior fossa floor, which is better than that obtained through a frontotemporal approach. In addition, the supraorbital approach decreases muscle pain and temporomandibular joint (TMJ) dysfunction from temporalis atrophy/injury that often follows a pterional craniotomy. The small incision needed for this technique heals faster, with less pain, and decreases hospital stay duration.

The major strength of the technique is in "width" of access and conversely, the weakness is in "height." Virtually any target area in both the ipsi- and contralateral anterior fossa can be reached, and when supplemented with endoscopy, even the

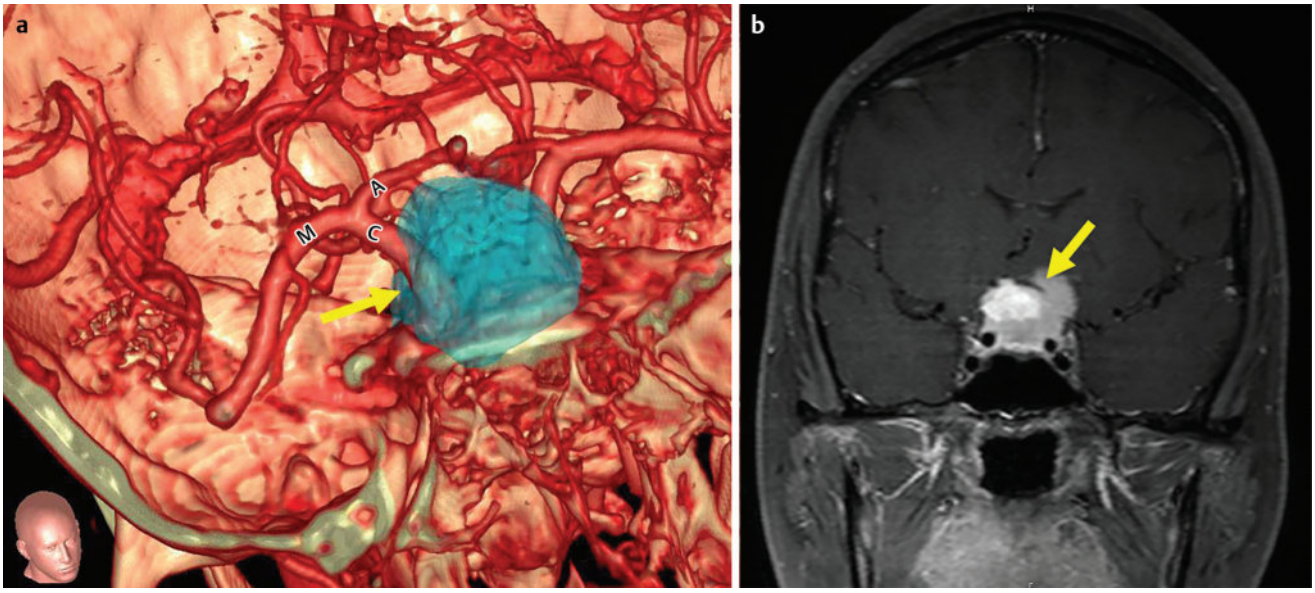


Fig. 1.2 Examples of encasement of major arteries by tuberculum sellae meningiomas. These meningiomas encase major arteries and are therefore riskier to remove via the endoscopic endonasal technique. The tumor obscures the arteries from the endoscope, and if injuries occur, arterial repair would be very challenging through the small endonasal opening. **(a)** Three-dimensional rendering of the tumor anatomy with the *yellow arrow* showing encasement of the right internal carotid artery at the supraclinoid segment. C, internal carotid artery; A, anterior cerebral artery; M, middle cerebral artery; **(b)** coronal MRI image of a different tumor with the *yellow arrow* showing the encasement of the left anterior cerebral artery in the A1 segment.

lateral cavernous sinus and ipsilateral retro-orbital space are accessible. The challenge for this approach is in reaching the top of lesions with significant superior extension. For tumors greater than 4 cm in height, an alternative approach should be sought.

While both eyelid and eyebrow incisions have been well documented in the literature for this approach, each carries a different set of advantages. The eyebrow incision is often utilized by neurosurgeons working alone without other expert assistance. It does carry risks of frontal paresthesia from supraorbital nerve injury, as well as eyebrow asymmetry from injury to the frontal branch of the facial nerve. The eyelid incision is usually performed under the guidance of oculoplastic surgeons, who are more familiar with the orbital soft tissue anatomy. The eyelid incision can be extended laterally without the same risks to the facial nerve branch, since it is made inferior to the course of the nerve. While both incisions heal quickly, the eyelid incision is naturally masked by the supratarsal crease and does not carry the risk of focal eyebrow alopecia.

Approach of Choice

While the cosmetic benefits of the eyebrow versus eyelid incision can be debated, at our institution, the specialized neuro-ophthalmology department has had extensive experience working with neurosurgeons in perfecting the eyelid approach exposure and closure to allow for maximum cosmetic advance. We find that this incision, coupled with the supraorbital craniotomy, provides the most direct access to anterior fossa lesions among transcranial approaches. As noted earlier, the main limitation of the technique is the vertical reach, but this is not a problem for our patient as her meningioma reached only up to 1.8 cm above the planum sphenoidale. Compared to other anterolateral transcranial approaches, the supraorbital craniotomy obviates any temporal lobe retraction while also allowing for early decompression of the optic nerve via drilling of the optic strut and release of the falciform ligament. For these above reasons, the transpalpebral, orbitofrontal craniotomy was our choice of approach for our patient's tumor, with the goal of safe maximal resection without additional neurological injury.

Questions

1. Which approach would *you* choose for this patient, and why?
2. What are the risks for catastrophic complications during your chosen approach? Vascular injury? Venous infarct? Cerebral edema?
3. How would you handle the dural closure for a supraorbital "keyhole" approach?

■ Description of the Technique

After establishment of general anesthesia, a lumbar drain was inserted prior to positioning to assist with brain relaxation and frontal lobe elevation. After that, the patient's head was positioned in the Mayfield head holder rotated about 15 degrees to the left with the neck slightly extended, putting the malar eminence at the highest point on the face. The right eyelid incision (**Fig. 1.3**) was made by our neuro-ophthalmology co-surgeons (see **The Three Approach Elements**).

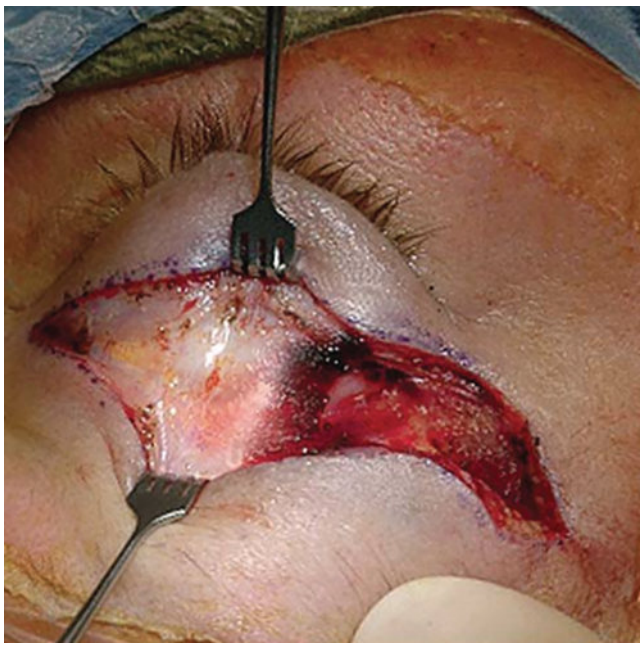


Fig. 1.3 Intraoperative image of the eyelid incision.

The Three Approach Elements

Corridor: anterolateral

Craniotomy: supraorbital "keyhole"

Modifier: anterior clinoidectomy

Important anatomical considerations for the exposure include the course of the frontal branch of the facial nerve, the supraorbital nerve, and the natural anatomical planes of the orbital septum. The lateral extension of the incision is limited to less than 2.5 cm lateral to the lateral canthus. The orbicularis oculi was divided carefully and dissected off of the orbital septum. Periosteal elevation was performed starting on the orbital side with attention toward preservation of the supraorbital neurovascular bundle.

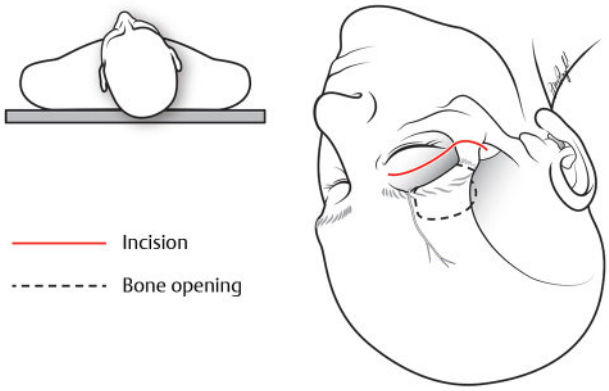
Operative Setup

Position: supine, head turned 15 degrees

Incision: transpalpebral

Bone opening: fronto-orbital "keyhole"

Durotomy: "upside down" U



Neuronavigation was used to plan a modified, one-piece, orbitofrontal craniotomy (**Fig. 1.4**). A spheno-orbital keyhole (the MacCarty burr hole) was made to access the frontal lobe dura and orbital periosteum. This burr hole was placed over the frontosphenoid suture about 1 cm posterior to the frontozygomatic junction. The frontal dura was exposed through the upper half of the hole, while the periorbita was exposed in the lower half. From the frontal portion of the burr hole, the craniotome was used to make the bone cut, initially heading superiorly 2 cm above and behind the supraorbital ridge, and then back again anteroinferiorly ending at the ridge, with care to finish *lateral* to the supraorbital foramen (notch) to avoid injury to the supraorbital nerve (**Fig. 1.5a**). After guarding of the orbital contents, the cut can be extended across the ridge with the craniotome. From the orbital half of the burr hole, a cut was made anteriorly across the zygomatic process of the frontal bone to

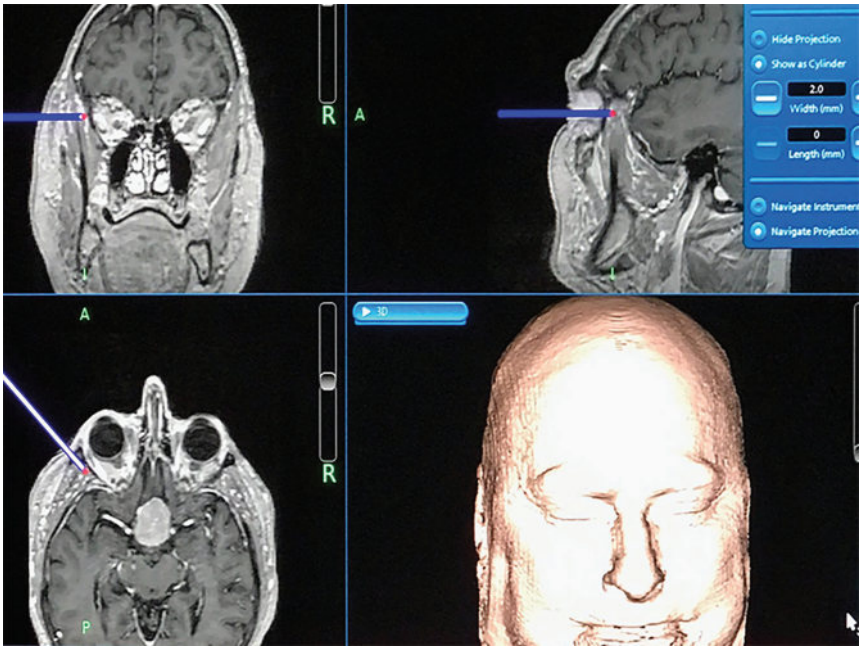


Fig. 1.4 Intraoperative localization with the neuronavigation system.

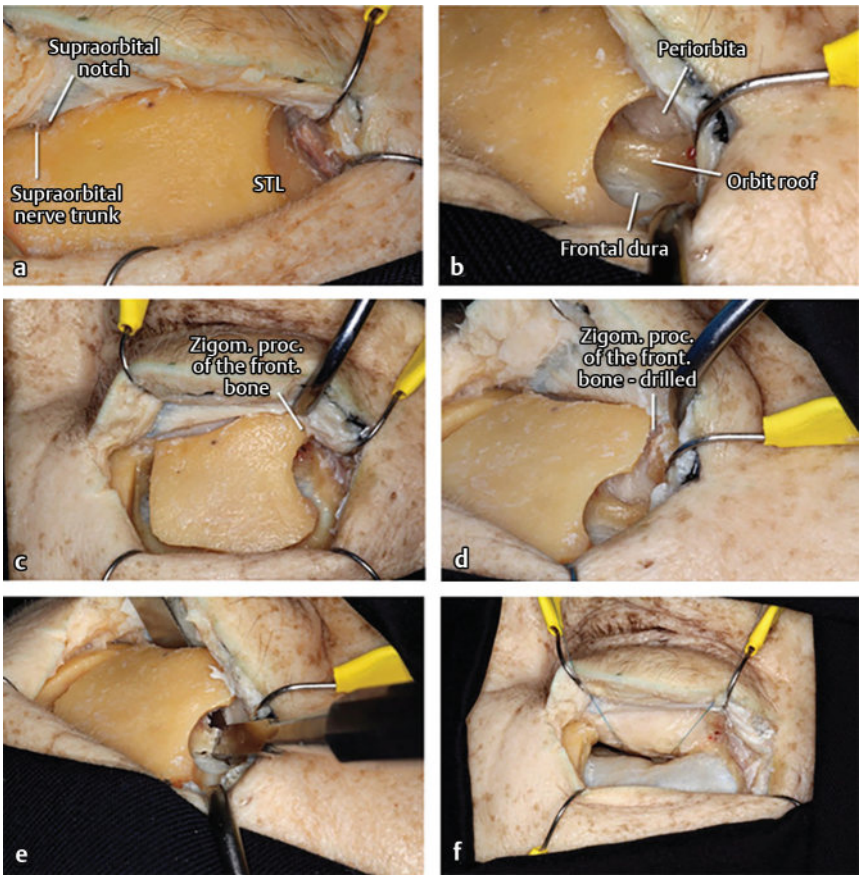


Fig. 1.5 Cadaveric dissection demonstrating the bone cuts of a fronto-orbital "keyhole" craniotomy. (a) An eyebrow incision was used in this dissection to demonstrate the craniotomy technique. Exposure was obtained lateral up to the superior temporal line and under the detached temporal muscle. **(b)** The MacCarty keyhole exposing the periorbita and the frontal dura separated by the orbital roof. **(c)** Osteotomies from the MacCarty keyhole following a U-shape line until the orbital rim lateral to the supraorbital notch. **(d)** Zygomatic process of the frontal bone drilled. **(e)** Osteotomes being used to cut through the orbital roof—note the protection of the periorbita with a spatula. **(f)** Sutures being used to retract the periorbita to gain more inferior space. STL, superior temporal line. (Reproduced from Mathias R, Lieber S, de Aguiar P, Maldaun M, Gardner P, Fernandez-Miranda J. Interfascial dissection for protection of the nerve branches to the frontalis muscles during supraorbital trans-eyebrow approach: an anatomical study and technical note. J Neurol Surg B Skull Base 2016;77 (3):265–270.)

detach the craniotomy from the lateral orbital wall (**Fig. 1.5d**). Finally, a small osteotome was used to cut across the orbital roof from the burr hole to the medial orbital roof cut, thus connecting all the bone cuts (**Fig. 1.5e**) (see **Operative Setup**).

Elevation of the frontal dura from the anterior fossa floor was accomplished with the assistance of the lumbar drain to relax the brain, and this opened the anterolateral subfrontal corridor. Using the drill under the microscope, the orbital roof was thinned and the bone overlying the right optic foramen was removed. This exposed the superior aspect of the optic nerve as it entered the orbit. Continuing the drilling lateral and then slightly inferior to the nerve, the optic strut was partially removed, and the extradural clinoidectomy was completed by detaching the anterior clinoid process at the optic foramen, optic strut, and the surrounding dura. Finally, some of the bone along the lateral wall of the superior orbital fissure was removed to increase working space and surgical freedom, and then the drilling was complete (**Fig. 1.6**).

The dura was opened in a curvilinear manner, and gentle, steady elevation of the frontal lobe with a brain ribbon opened the subfrontal corridor inside the dura (**Fig. 1.7a**). The tumor was devascularized from its blood supply along the floor of the anterior fossa as far as the corridor permitted and then the tumor was debulked internally (**Fig. 1.7b**). This maneuver yielded more space, and this was exploited to dissect the tumor capsule away from the neurovascular structures nearby. The ICA branches and optic nerves laterally on both sides, ACAs superiorly, and the optic chiasm posteriorly, were all detached from the tumor capsule (**Fig. 1.7c**). After this, the rest of the tumor was removed completely (**Fig. 1.7d**), thus achieving the surgical goal we had determined at the outset of the operation (**Fig. 1.8**).

After the intracranial portion of the case was complete, the dura was closed in a watertight fashion to prevent postoperative pseudomeningocele, which can significantly delay both wound healing and hospital discharge. The bone flap was replaced and secured with two to three low-profile titanium plates. Multiple holes were made in the temporal bone and zygomatic process of the frontal bone with a B5 tack-up bit to allow adequate suture reapproximation of the temporalis muscle. Our ophthalmology colleagues completed the closure by placing a constant-pressure, closed-suction (TLS) system to assist wound healing. The incision was closed using 4–0 absorbable Vicryl sutures followed by 6–0 running sutures for the skin.

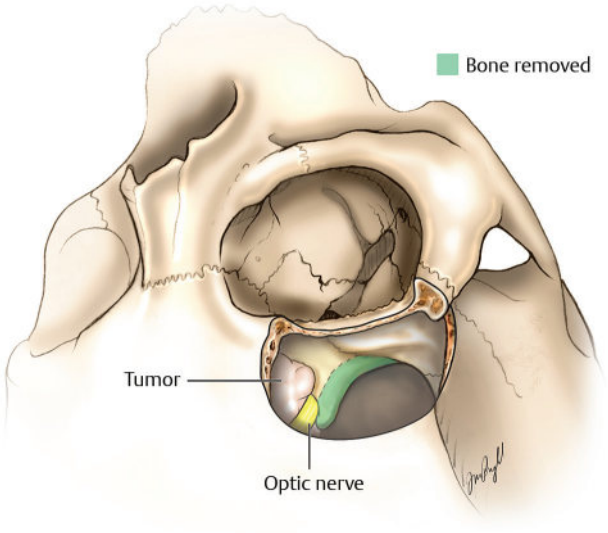


Fig. 1.6 The fronto-orbital craniotomy widely exposes the floor of the anterior fossa.

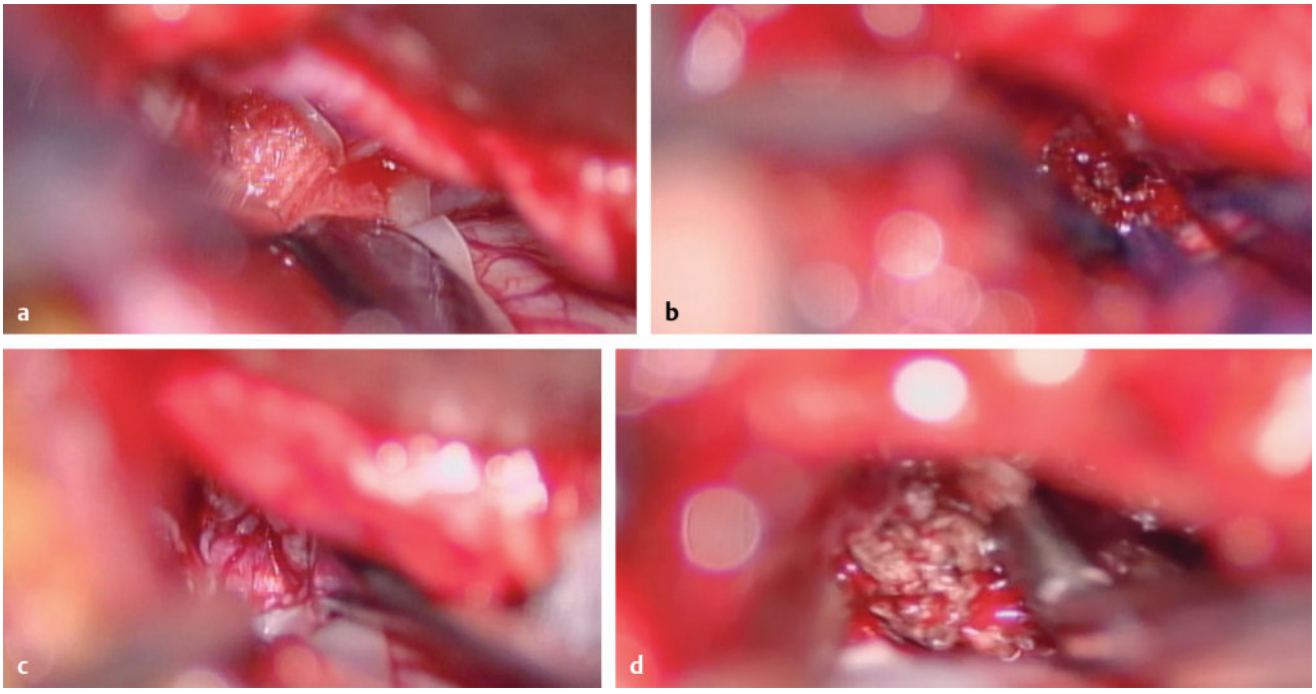


Fig. 1.7 Intraoperative image under the microscope. **(a)** The tumor capsule came into view in the subfrontal corridor. **(b)** The tumor was internally debulked using the suction–cautery technique. **(c)** Sharp dissection was used to detach critical neurological structures away from the tumor capsule. **(d)** The final piece of the tumor was lifted out of the resection cavity with pituitary rongeurs.

Surgical Pearls

1. A well-trained oculoplastic/neuro-ophthalmology co-surgeon can be essential to a smoothly functioning team that will provide the best cosmetic outcomes for the patient in the long run.
2. Large anterior fossa lesions with significant lateral extension into the middle fossa, or significant superior extension above the plane of the anterior fossa floor must be considered carefully on preoperative imaging before choosing an orbitofrontal approach. For such lesions, potential neurovascular complications can occur during dissection of the lateral and superior margins of the lesion.
3. Lumbar drain placement preoperatively facilitates frontal lobe retraction and minimizes need for fixed retractors.
4. Watertight dural closure and postoperative use of the drain decreases the risk of pseudomeningocele formation that could lead to a poor cosmetic result given the thin skin of the region.

Aftercare

Our patient was monitored in the neurosurgical intensive care unit (ICU) on the first night after surgery. A computed tomography (CT) scan confirmed our intraoperative assessment of a complete resection (**Fig. 1.9**). She was followed by the endocrinology team for potential pituitary-related postoperative issues. Her lumbar drain was gradually weaned off over the following days to decrease the risk of pseudomeningocele formation, and was removed after 5 days. Her constant pressure TLS drain was removed by the ophthalmology team on the second day after surgery. She was discharged to inpatient rehabilitation on postoperative day 6 continuing on twice-daily oral steroids under endocrinology management.

By her second week follow-up, her left-sided temporal visual field deficits had essentially returned to baseline with noted improvements in the right peripheral visual fields as well. At the 2-month follow-up, the patient's incision was well-healed and all residual periorbital edema had resolved with an excellent cosmetic result (**Fig. 1.10**). She was cleared to return to

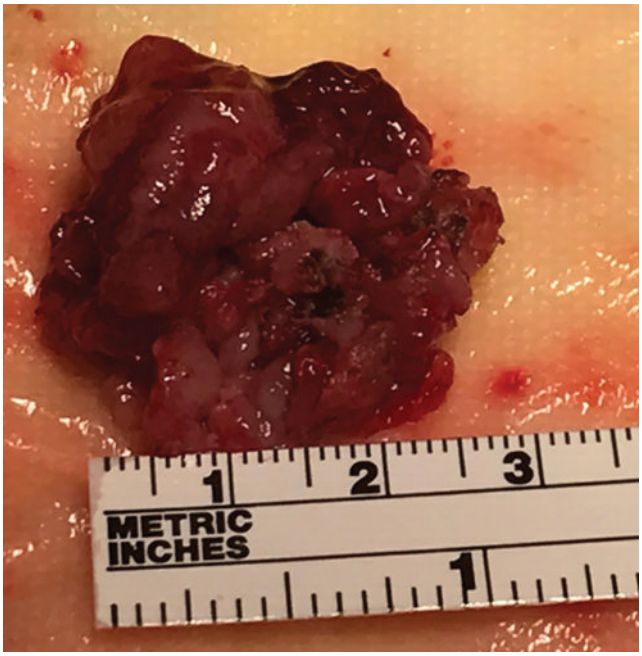


Fig. 1.8 The surgical specimen after complete tumor resection.

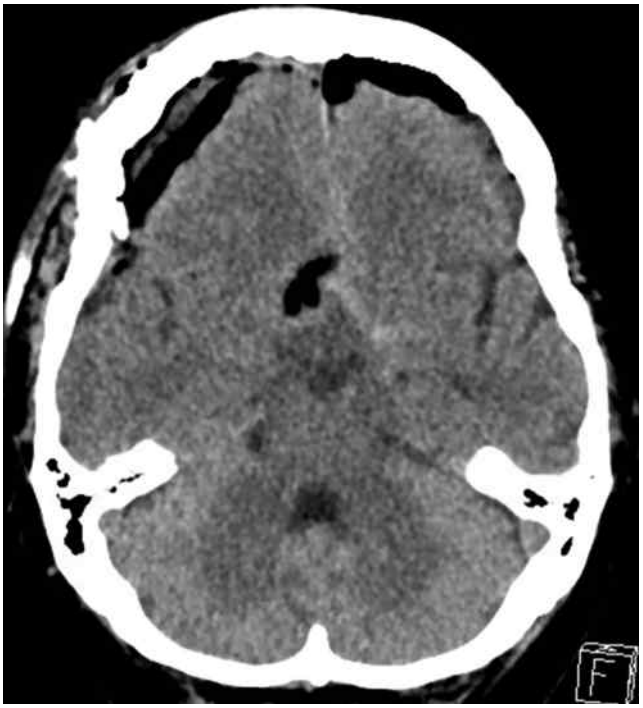


Fig. 1.9 Postoperative CT head scan after the tumor resection.

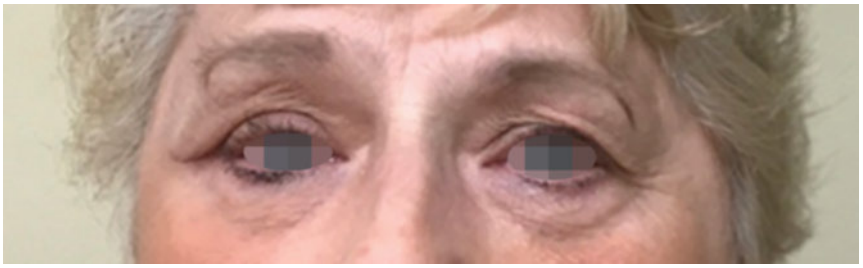


Fig. 1.10 **Two-months follow-up in clinic.** The incision was well-healed and the eyelids were essentially indistinguishable between right and left side.

full-time teaching duties. Final pathology showed a transitional type WHO grade 1 meningioma. No adjuvant chemotherapy or radiation treatment was offered.

■ Possible Complications and Associated Management

The transpalpebral approach carries a small risk of postoperative ptosis, as well as double or blurry vision from globe retraction. Rare cases of postoperative ptosis improve naturally on their own after 8 to 12 weeks, and is usually partially related to residual periorbital edema at the surgical site. In rare, persistent cases that affect vision or cause significant facial asymmetry, oculoplastic surgeons can perform blepharoplasty that restores eyelid positioning. Visual symptoms such as blurry vision and double vision are rare, but fortunately, these resolved without intervention as well. Our patients continue to follow up with ophthalmology for the first 6 to 12 months after their operation.

Perspective

Hermes G. Garcia and James J. Evans

■ Introduction

Over the last decade, the optimal surgical approach for symptomatic tuberculum sellae and planum sphenoidale meningiomas has stimulated a great deal of debate. Transcranial microsurgical resection, whether through pterional, orbitozygomatic, or bifrontal approach, had been the standard treatment for these tumors, but they all require frontal lobe retraction with its inherent neurological risk. Keyhole craniotomies, such as eyebrow or eyelid supraorbital approaches detailed in the first section of this chapter, have been advocated for better cosmesis. Endoscopic endonasal approaches (EEAs) offer an additional option, which provides direct access to the dural attachment of the tumor, with similar surgical results compared to the open approaches. The modern cranial base surgeon should have all of these options at his/her disposal, and with that, pairing a specific patient with the optimal approach becomes a critical decision for the best clinical outcome.

There are several advantages of the EEA over transcranial procedures. As an inherent part of the approach, the EEA requires removal of the tuberculum sellae and/or planum sphenoidale in order to access the tumor. This improves the Simpson grade of resection, and decreases the likelihood for recurrence especially in cases where the dura and bone of the skull base are infiltrated with tumor. Likewise, early coagulation of the dural attachment eliminates the feeding vessels to the tumor *before* the resection. This results in less blood loss and a cleaner surgical field in which to dissect the tumor–arachnoid interface. The EEA also provides a direct route to the tumor without brain retraction. This may decrease the risk of frontal lobe dysfunction and postoperative seizures. In addition, endoscopic visualization provides an unparalleled view of the microvasculature of the inferomedial surface of the optic nerves and chiasm. This affords the surgeon an opportunity to dissect these structures meticulously for preservation, which may account for the improved visual outcomes reported for EEA.

The EEA also has certain disadvantages that need to be considered. There is a steep learning curve with EEA, requiring a progressive increase in the complexity of endonasal cases prior to attempting resection of intradural tumors. Also, the removal of the bone and dura of the skull base with the endonasal approaches creates a sizable dural defect necessitating careful repair. This accounts for the increased risk of postoperative cerebrospinal fluid (CSF) leak with these approaches. Surgical access may be limited in areas lateral to the tuberculum sellae, and specifically, tumor extensions lateral to the optic nerves and internal carotid arteries are challenging to reach through the endonasal corridor. Finally, vascular encasement increases the risk of complications for both transcranial and endoscopic endonasal approaches; however, management of vascular injuries is more difficult in endonasal approaches.

Some of these disadvantages can be circumvented with appropriate training, case selection, surgical goal consideration, and experience with endonasal reconstruction, as the following discussion highlights.

■ Case Presentation

A 50-year-old right-handed woman presented with diminished vision in the left eye. She was evaluated by an ophthalmologist and found to have significant visual loss in all quadrants of the left eye. More specifically, she had mild optic nerve pallor with visual acuity 20/20 OD and 20/40 OS. MRI revealed an extra-axial enhancing tumor based at the tuberculum sellae with extension into the sella, most consistent with an anterior cranial base meningioma (**Fig. 1.11**). Her endocrine/pituitary review of systems and bloodwork were negative.

■ Anatomical and Therapeutic Considerations

The patient's tumor compressed the optic nerves bilaterally as well as the optic chiasm. The optic nerves were displaced laterally, but there was no extension of tumor lateral to the optic nerves. Similarly, the tumor did not extend laterally beyond the internal carotid arteries. Superiorly, it extended up to the anterior cerebral arteries, which were displaced superiorly. Very importantly, the anterior cerebral arteries were not encased by the tumor. The CT angiogram verified this anatomy and showed no evidence of calcification within the tumor (**Fig. 1.11, Fig. 1.12, Video 1.1**).

In our experience, there are few absolute contraindications for the EEA for tuberculum sellae meningiomas (**Table 1.1**). However, there are certain anatomical and radiographic factors which must be thoroughly assessed on preoperative imaging to determine if EEA is appropriate. For example, the relationship of the tumor to the optic nerves should be carefully evaluated. Tumors that are located medial and inferior to the optic nerves are better approached through an EEA than a transcranial approach, while tumor lateral to the optic nerve cannot be safely accessed through an endonasal approach. Although tumor superior to the optic nerve can be reached by the EEA, there is limited visualization of the superior surface of the nerve and its microvasculature via this approach. For this reason, tumor in this location is better addressed via a transcranial approach. We would not recommend an endonasal approach with attempted gross total resection, if there were significant extension of tumor superior and particularly lateral to the optic nerves.

The position of the chiasm relative to the tumor should also be carefully analyzed. A prefixed chiasm would obstruct the surgical corridor from a transcranial route, and therefore an EEA may be favored in these cases. Tumors with significant sella extension could be accessed via either route, but this is more directly approached with an EEA. If there is significant ventral extension of the tumor toward the cribriform plate in the setting of normal olfaction, one might consider a transcranial approach to better preserve olfaction. Frontal lobe edema suggesting pial invasion is not a contraindication for an endonasal approach, as safe bimanual dissection can be done in a similar fashion to a transcranial approach. However, a large and heavily calcified

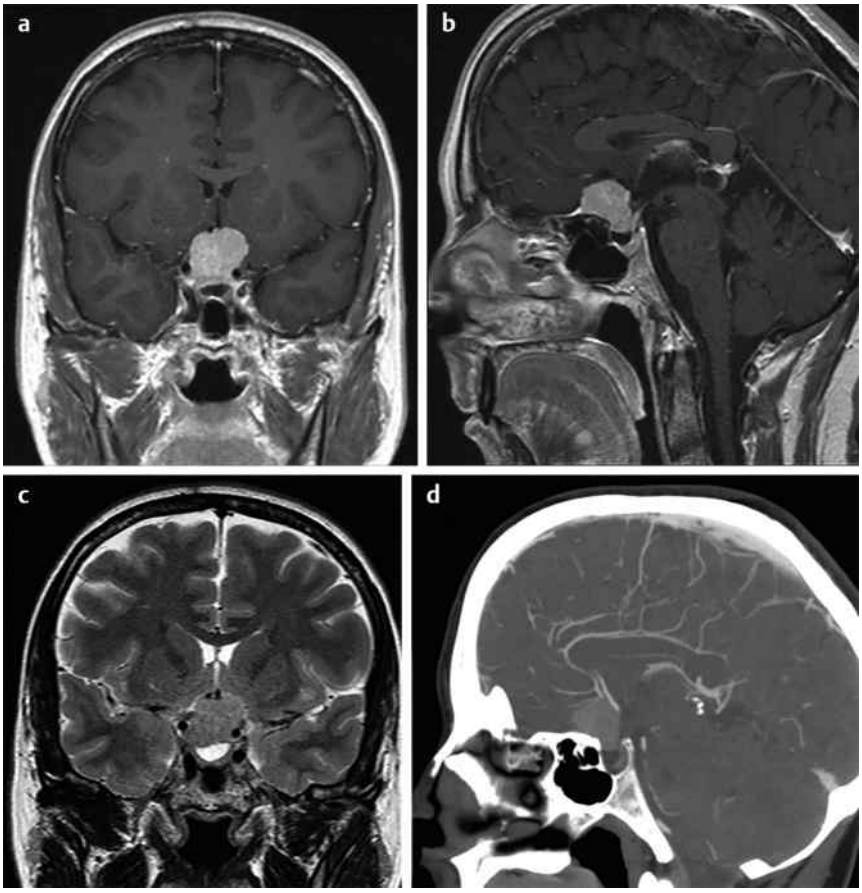


Fig. 1.11 Preoperative imaging. **(a)** Coronal T1 postcontrast sequence MRI showing the suprasellar extension of the tuberculum sellae meningioma. The optic nerves were displaced laterally and there was no extension of tumor lateral to the optic nerves or internal carotid arteries. **(b)** Sagittal T1 postcontrast sequence MRI showing the origin of the tumor along the planum and tuberculum sellae. There was extension into the sella with displacement of the infundibulum posteriorly. The dural tail and mass did not extend anteriorly beyond the planum. **(c)** T2 coronal MRI sequence showing the ACAs draped on the superior surface. The tumor was in contact with approximately 180 degrees of the surface of these. Although there was no obvious CSF cuff, lack of 360-degree encasement makes this tumor amenable for endoscopic endonasal resection. **(d)** Sagittal CT angiogram showed lack of extensive calcification. The ACAs were draped superior and posterior with direct branches emanating on the superior surface of the tumor, yet not embedded within the tumor.

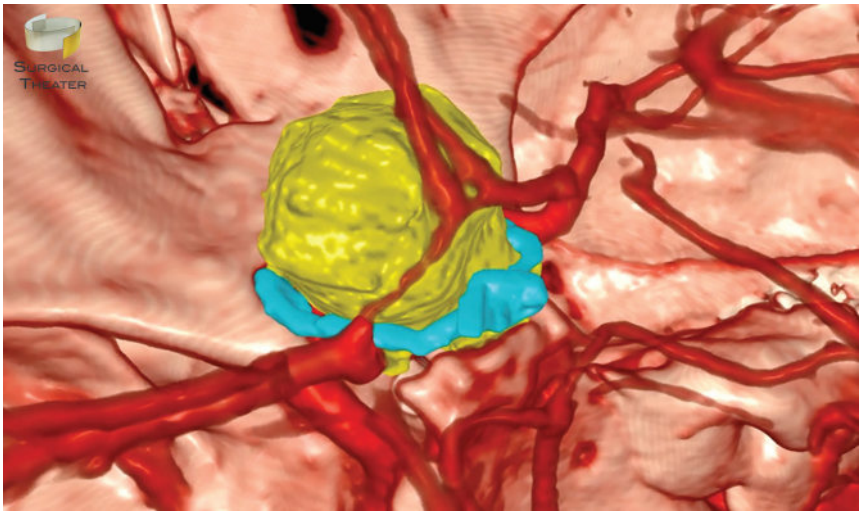


Fig. 1.12 Three-dimensional rendering of the tumor anatomy of the patient's tuberculum sellae meningioma. The view is posterior-to-anterior, left-to-right. This demonstrates that the tumor displaced the neurovascular structures posteriorly and superiorly. The left A1 is most affected, but the relationship with the tumor was short of encasement. The optic nerves were displaced laterally and there was no extension of tumor lateral to the optic nerves or internal carotid arteries.

tumor may be more difficult and time consuming to manipulate from an endonasal route and thus may present as a relative contraindication for EEA.

The vascular anatomy around the tumor is always an important determinant in selecting an approach. A very narrow intercarotid distance ("kissing carotids") will obstruct safe access to the tumor, and therefore would be another contraindication for an EEA (**Fig. 1.13**). Vascular encasement of the ICA and ACAs needs to be considered prior to attempting an EEA. Encasement can certainly limit the degree of resection of the tumor and increase the risk of vascular injury. Despite the advances in endoscopic endonasal techniques and instrumentation, a reliable method for direct repair or reanastomosis of vessels has not yet been developed. The degree of adhesion between a vessel and the tumor is hard to assess radiographically; however, there are findings that may provide hints. Tumor in contact with the cavernous segment of the ICA, proximal to the distal dural ring, is more likely to invade the adventitia of the ICA, thus increasing the risk of ICA injury. Prior radiation and surgery will also likely obliterate the arachnoid plane at the tumor–vessel interface,

and therefore in these cases, an EEA may not be advisable. Partial encasement of the ACAs with evidence of a thin spinal fluid cuff is amenable to either transcranial or endonasal approach. This is best evaluated on thin-cut T2 coronal imaging, where evidence of circumferential 360-degree encasement may favor a transcranial approach.

Table 1.1 Anatomical and radiographic factors to evaluate for choosing the optimal surgical approach

Endonasal	Transcranial
Absolute indication	
Sella extension	Extension lateral to the optic nerve
	ACA encasement
	Kissing carotids
Relative indication	
Tumor inferior to ipsilateral optic nerve	Abuts ACA with history of radiation
	Ventral extension to cribriform
	Evidence of no viable nasoseptal flap

Abbreviation: ACA, anterior cerebral artery.
Source: Adapted from Kshettry VR, Elshazly K, Evans JJ. Endoscopic transnasal surgery for planum and tuberculum sellae meningiomas: decision-making, technique and outcomes. CNS Oncol 2016;5 (4):211–222.

While the factors listed above describe the anatomical and radiographic factors to consider when choosing an approach, the surgical goal should always serve as the ultimate guide. Safe, maximal tumor resection should always be considered; however, for complex tuberculum sellae tumors where a gross total resection is not feasible, one should choose the approach that best addresses the specific surgical goal. In most cases, the limited goal involves decompression of a specific structure, such as the chiasm or optic nerve, and the approach that best targets that structure is often the best option.

Regarding the case presented above, there are multiple factors favoring an EEA. The patient was young and without significant medical comorbidities; therefore, a safe maximal resection with the goal of gross total resection was planned. The tumor compressed the inferomedial aspect of the optic canals bilaterally. This displaced the optic nerves laterally favoring an endonasal approach. From a vascular standpoint, there were no kissing carotids, no extension lateral to the ICAs, and the ACAs were not circumferentially encased. Review of the CT did not show heavy calcification and there was no significant ventral extension toward the cribriform. There were also no anatomical factors or prior sinonasal procedures that would limit an adequate endonasal cranial base reconstruction. We therefore recommended an EEA for decompression of the optic system and attempted gross total resection of the tumor.

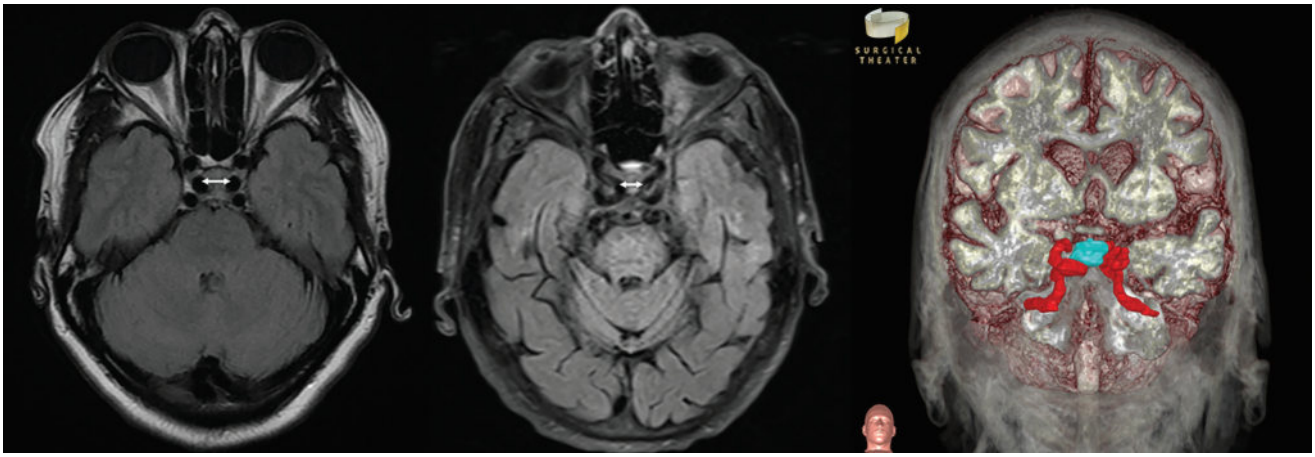


Fig. 1.13 "Kissing" carotids. MRI and three-dimensional rendering of "kissing" carotids with small intercarotid distances (*white arrows*). The narrow gap between the carotid arteries blocks the surgical exposure of the endonasal endoscopic approach (EEA) to areas around the sella turcica. It is therefore a contraindication for EEA.

Description of the Technique

After general endotracheal anesthesia, the patient was placed supine, 180 degrees from the anesthesiologist, and with the head placed on a circular foam headrest. A stereotactic navigation mask was used without skull fixation. The neurosurgeon and otolaryngologist stood on opposite sides of the patient with dual video and navigation monitors placed opposite of each surgeon. As the EEA is a clean contaminated surgical procedure, no specific skin or nasal surgical prep was used. We do not use lumbar drains. Ancef was administered within 1 hour prior to surgical incision and 10 mg of dexamethasone was given due to significant tumor compression of the optic nerves and chiasm.

The nasal cavity was prepared with oxymetazoline-soaked cottonoid pledgets placed in each nasal cavity and 1% lidocaine with 1:100,000 epinephrine was injected into the nasal mucosa. The middle and inferior turbinates were lateralized bilaterally. We do not routinely remove the middle turbinates, as we have found that it does not increase the access to the sella and particularly midline lesions. A pedicled nasoseptal flap was harvested. Wide bilateral sphenoidotomies were performed with care to preserve the pedicle of the nasoseptal flap. Bilateral posterior ethmoidectomies were then performed to allow exposure from planum to the sella. A small posterior septectomy (< 15 mm) was made for binaural access. The mucosa of the sphenoid sinus was stripped to avoid postoperative mucocele formation beneath the final nasoseptal flap repair. All sphenoid sinus septations were drilled down to eliminate dead space between the nasoseptal flap and bone during the reconstruction after tumor resection.

Once inside the sphenoid, the key anatomical landmarks were identified, including the planum sphenoidale, tuberculum sellae, sella, lateral optic–carotid recesses (OCRs), optic canals, and clinoidal carotid protuberances (**Fig. 1.14, Fig. 1.15a**). The bone from mid-to-upper sella was removed with a combination of diamond drill and the Kerrison punches. Next, the bone of the tuberculum sellae and planum was removed. This bone is often hyperostotic and can be vascular due to the adjacent tumor fed by the ethmoidal arteries. The use of a diamond burr is safer than a cutting burr when working around critical neurovascular structures and is also helpful for bony hemostasis. We use intraoperative neuronavigation to tailor the drilling of the planum sphenoidale, sufficient to remove a margin of dura around the anterior extent of the tumor (**Fig. 1.15b**). The bone overlying the proximal optic canals was unroofed to access the tumor extension into the medial optic canals bilaterally. Drilling with continuous irrigation was performed to avoid thermal injury to the optic nerves. The lateral OCRs help estimate the location of the canalicular segment of the optic nerve. A key to unlocking wide access to the optic–carotid cistern is removal of the lateral strut of the tuberculum sellae. In sphenoid sinuses with limited aeration and anatomical landmarks, neuronavigation

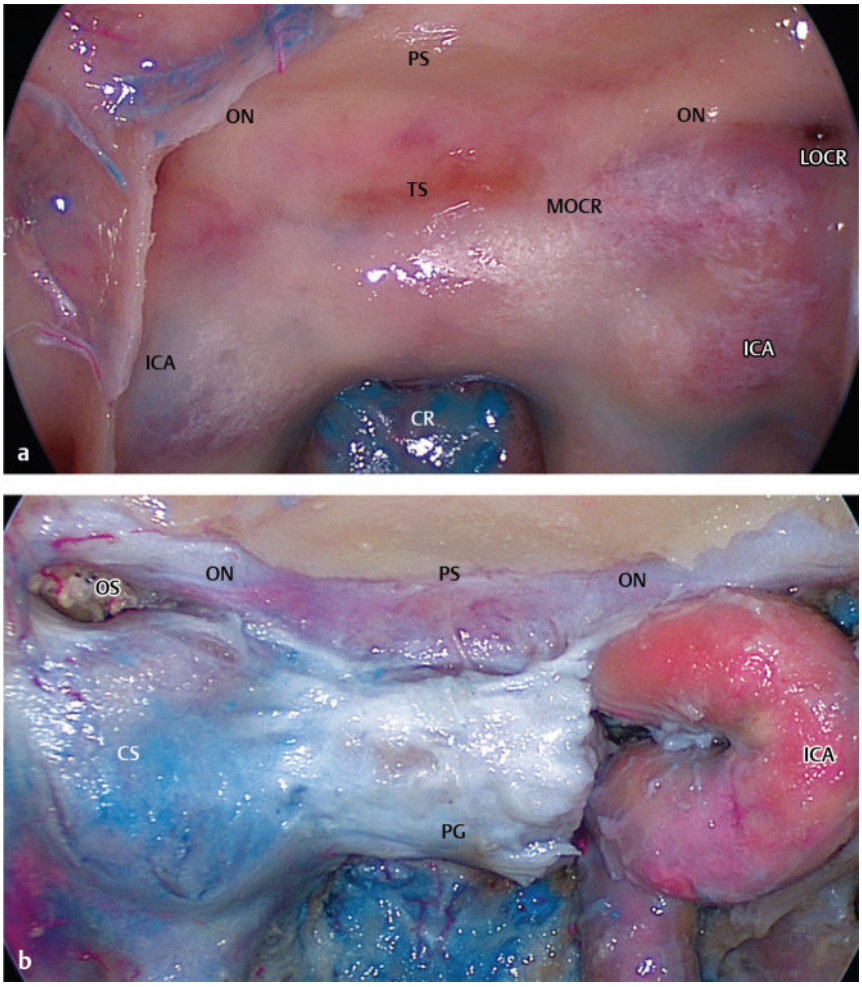


Fig. 1.14 Cadaveric dissection. (a) Exposure and identification of the anatomical landmarks during the endoscopic endonasal approach. **(b)** After bone removal, the underlying structures and their anatomical relationships with the optic nerve are appreciated. CR, clival recess; CS: cavernous sinus; ICA, internal carotid artery; LOCR, lateral optic–carotid recess; MOCR, medial optic–carotid recess; ON, optic nerve; OS, optic strut; PG, pituitary gland; PS, planum sphenoidale; TS, tuberculum sellae. (Reproduced from Beer-Furlan A, Evins A, Rigante L, Anichini G, Stieg PE, Bernardo A. The pterional port in dual-port endoscopy: a 2D and 3D cadaveric study. J Neurol Surg B Skull Base 2015;76 (1):80–86.)

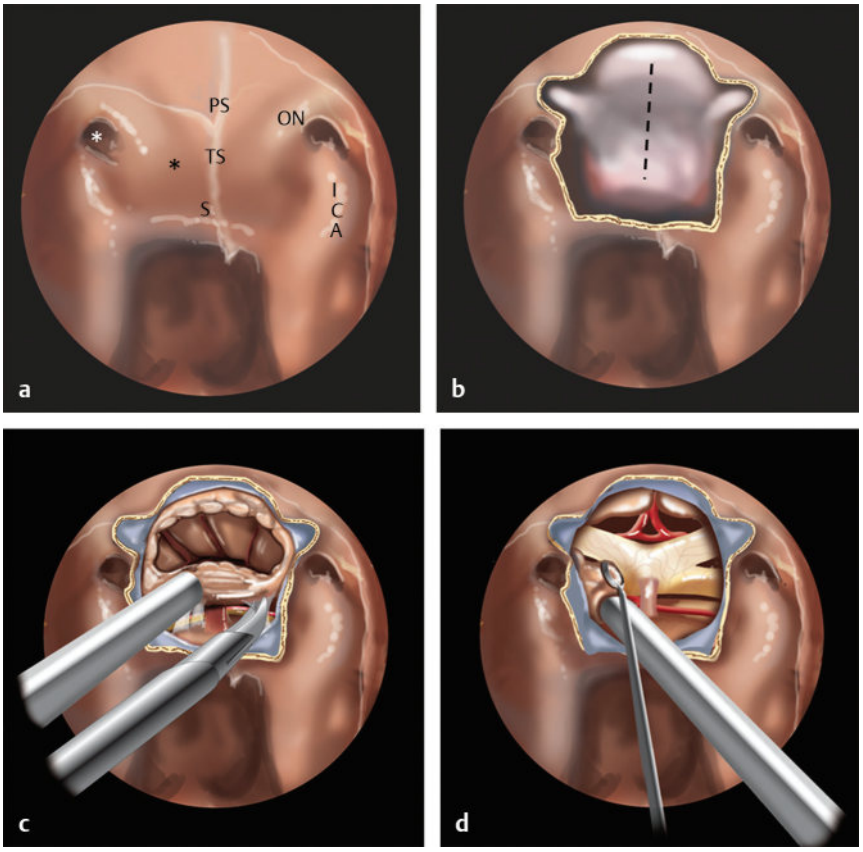


Fig. 1.15 Operative exposure and tumor resection. (a) Upon completion of the nasoseptal flap harvest and sphenoidotomy, the location of the key operative landmarks should be identified: sella (S), tuberculum sellae (TS), planum sphenoidale (PS), and the paired cavernous internal carotid arteries (ICAs), optic nerves (ON), and the lateral optic–carotid recesses (*white asterisk*). The lateral tubercular struts (*dark asterisk*) were removed to improve maneuverability of instruments. **(b)** The osteotomy was completed. Removal included the bone overlying the sella, and anteriorly to include portions of the planum sphenoidale sufficient to expose the dural base of the tumor. After extensive coagulation of the dura underlying the tumor, a linear vertical incision was done to start the internal debulking (*dashed line*). **(c)** After aggressive internal debulking, the tumor was dissected from the surrounding neurovascular structures with use of microdissection. **(d)** Last remnant of tumor, which was attached to the adventitia of the ICA, was gently removed. A small remnant was left but carefully coagulated.

and micro-Doppler are useful adjuncts to determine the location of the optic nerve and the carotid artery.

Once exposure was completed, we proceeded with the resection of the tumor. Endonasal meningioma resection follows the same general principles of microsurgical meningioma resection with generous coagulation of the dura to disrupt blood supply to the meningioma. An initial dural opening limited to the central aspect of the tumor was done and internal debulking was achieved with a combination of suction, micro-scissors, and ring curettes (**Fig. 1.15c**). This initial dural opening preserved the tumor attachment to the dura, which can limit transmission of dissection forces to surrounding critical neurological structures, such as the optic system, during the debulking.

After internal debulking, the dura was then incised circumferentially around the base of tumor. Using bimanual sharp and blunt extracapsular arachnoid dissection under direct endoscopic visualization, the remainder of the tumor was removed in large blocks. During the dissection of the optic nerves and their blood supply, the anterior communicating arteries, and posterior communicating arteries, one should always use bimanual sharp microdissection. In our case, careful dissection of the arachnoid plane between the ACAs and the tumor facilitated gross total resection. There was an area of dural attachment along the adventitia of the right ICA near the distal dural ring that could not be entirely resected. However, it was carefully cauterized with concentric suction bipolar electrocautery (**Fig. 1.15d, Fig. 1.16**).

Once the removal of the tumor was complete, we proceeded with the reconstruction of the dural defect (**Fig. 1.17**). Among centers performing this type of surgery, there are significant variations in techniques and materials used for dural reconstruction. A common theme, however, is a stable primary dural repair followed by a pedicle vascularized mucosal flap. For the primary dural repair, we used a fascia lata bilayer inlay/onlay "button" graft. The size of the dural defect was measured using the tips of a pituitary rongeur as a caliper, and an appropriately sized button graft was made. The two layers of fascia lata were sutured together using 4–0 Nurolon suture (**Fig. 1.17a**). This helps to hold both layers in close approximation to the dura and doubles the surface area for healing. Once in adequate position, the graft transmitted the normal dural pulsations without evidence of CSF leakage indicating that a good seal had been achieved. The nasoseptal flap was then placed over the dural reconstruction and surrounding sphenoid bone with care to ensure that there is no

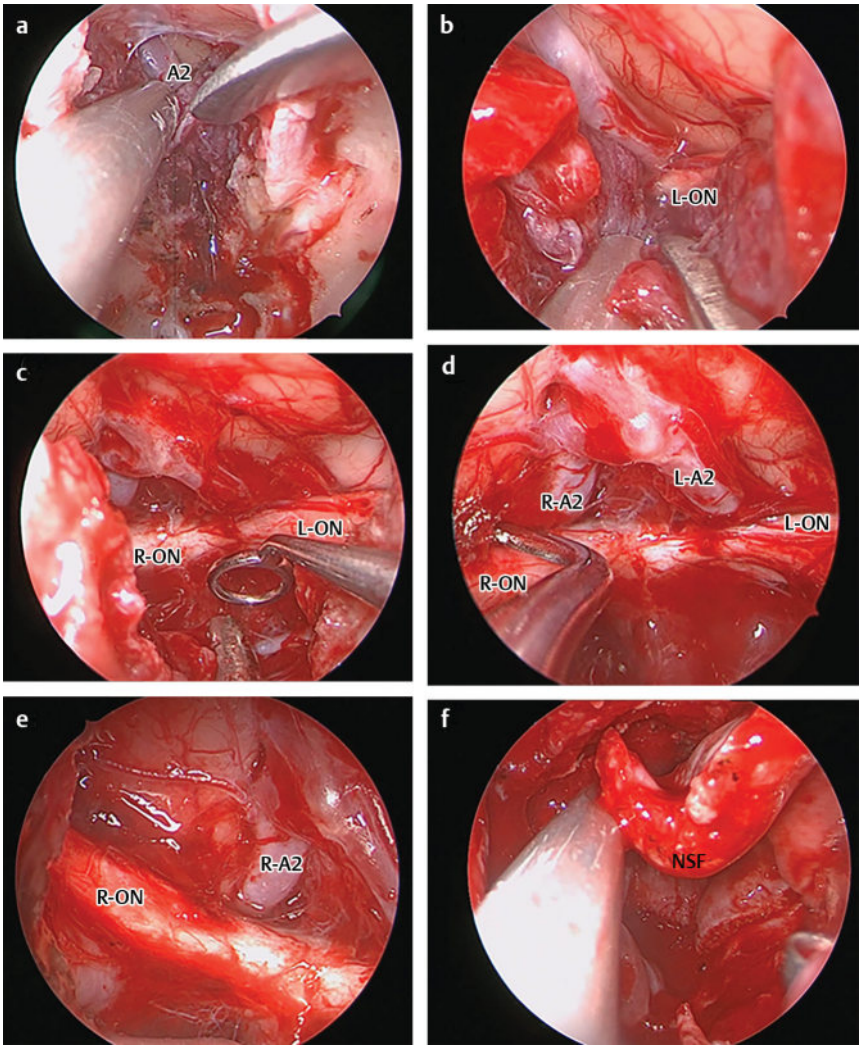


Fig. 1.16 Resection of a tuberculum sellae meningioma via EEA. The techniques described in the text are demonstrated here in a separate patient with a similar tumor. **(a)** After a central dural opening, the tumor was dissected from the left A2 segment of the anterior cerebral artery. **(b)** Next, the left optic nerve was dissected away from tumor. **(c)** After freeing both optic nerves, the tumor was removed from around the optic chiasm. **(d)** Although tumor superior to the optic nerve is a relative contraindication for EEA, here a tiny tumor component above the right optic nerve was dissected and removed. **(e)** Final inspection of the surgical field with an angled endoscope. **(f)** Placement of the nasoseptal flap for closure. L-A2, left A2; L-ON, left optic nerve; NSF, nasoseptal flap; R-A2, right A2 segment of the anterior cerebral artery; R-ON, right optic nerve. (Photos courtesy of Drs. A. Anaizi and T. Deklotz.)

dead space between the flap and bone. Polyethylene glycol (PEG) hydrogel glue was applied to secure the edges of the nasoseptal flap. The middle and inferior turbinates were then medialized. Absorbable nasal packing is placed in the middle meatus bilaterally to keep the middle turbinates medialized.

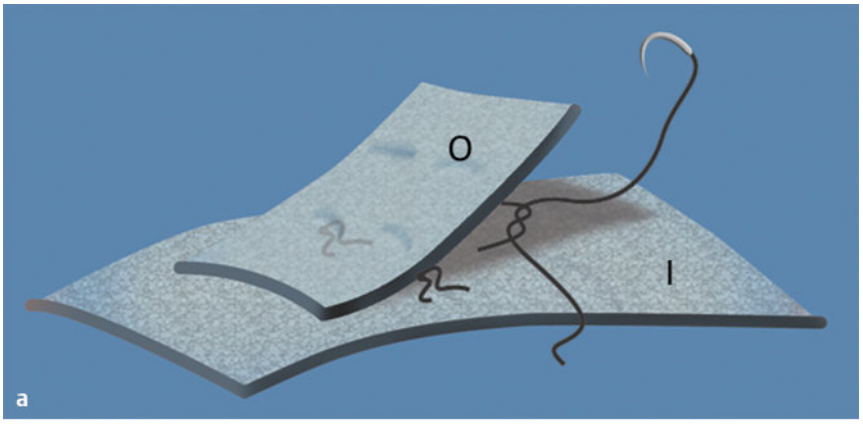
■ Aftercare

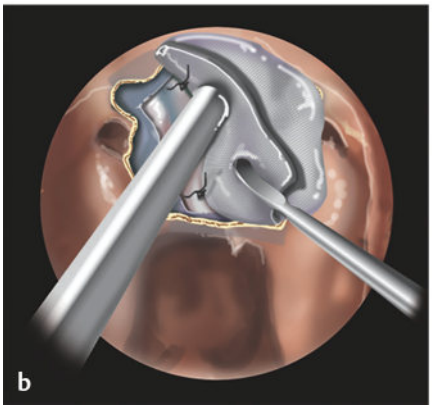
Postoperatively in the ICU, the patient was kept with the head of bed above 45 degrees for the first 48 to 72 hours after surgery. This diminishes bloody nasal oozing, edema of the sinonasal mucosa, and the risk of postoperative CSF leakage. Our patient's recovery was unremarkable, and her endocrine profile remained normal. The latest eye examination was normal with 20/20 acuity bilaterally and full visual fields. Follow-up MRIs have revealed excellent resection without evidence of residual tumor (**Fig. 1.18**).

■ Commentary

The EEA offers several advantages over a transcranial approach in select cases. In this discussion, we highlighted the nuances regarding case selection as well as intraoperative maneuvers for safe and effective surgery. In this particular patient with the anatomical details previously described, we were able to achieve an excellent tumor resection and the patient's vision improved back to normal with preservation of endocrine function.

It is important to emphasize that the endonasal resection of the tumor is performed with standard bimanual sharp and blunt microdissection techniques similar to open transcranial approaches. We recognize, however, that there is a steep learning curve for expanded EEAs. These procedures should be attempted only after the entire team has gained experience with less complex endoscopic endonasal procedures. One must





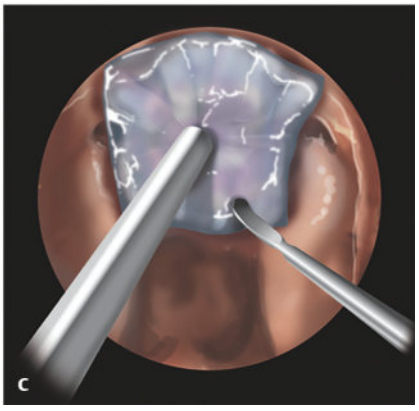


Fig. 1.17 Cranial base reconstruction. **(a)** Fascia lata was harvested and the size of the button graft was appropriately made after measuring the cranial base defect. The inlay (I) should be larger than the onlay (O) and they are sutured together with four knots. **(b)** The button graft was fashioned carefully with use of suction and dissectors, so the inlay covered the dural defect internally and the onlay covered the defect in contact with the dura. Once in place, we circumferentially inspected the borders under the inlay to confirm adequate placement. **(c)** Lack of CSF leak and the transmission of normal dural pulsations also confirmed adequate placement. The nasoseptal flap was placed over this graft.

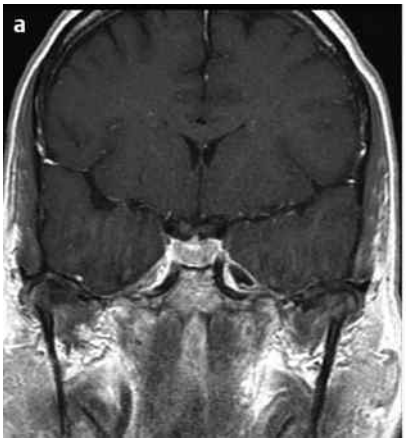


Fig. 1.18 One-year postoperative imaging. **(a)** Coronal and **(b)** sagittal T1 postcontrast sequence MRI revealing no residual tumor for good decompression of the optic apparatus. In **(b)**, there is good viability of the nasoseptal flap, which is evidenced by the avidly enhancing mucosa along the planum and rest of sella (*).

also keep in mind that the skull base repair is just as important as the approach and tumor removal. With a good algorithm of techniques for cranial base repair, the rate of postoperative CSF leak can be minimized and should not deter the surgeons from utilizing this approach.

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2 Olfactory Groove

Angela E. Downes and A. Samy Youssef

Keywords: olfaction, cribriform plate, endoscopic, transcribriform approach, transbasal approach

■ Case Presentation

A 55-year-old female, with a history of Lyme disease, underwent work-up for her headaches, and was found to have a 16-mm mass on the anterior skull base. This was deemed too small to be the cause of her headaches. Two years later, she presented to clinic with constant right frontal headaches, and a repeat magnetic resonance imaging (MRI) showed enlargement of the contrast-enhancing mass, which measured 24 × 26 mm at that point. The tumor was located at the cribriform plate, and originated from the right side (**Fig. 2.1**). She reported decreased sense of smell and taste, as well as a progressive dysosmia, whereby she experienced distorted smell of familiar substances.

Questions

1. How would you manage this lesion? Observation? Radiation? Or surgery?
2. How does the olfactory function affect the next step in management?
3. What is the objective assessment for olfaction?
4. If surgery is pursued, what would be your surgical approach?

■ Diagnosis and Assessment

The MRI showed a homogenously contrast-enhancing, extra-axial mass originating from the right cribriform plate of the ethmoid bone without surrounding edema. The tumor was anterior to the optic chiasm, anterior to the cerebral arteries and pituitary gland, and did not invade the ethmoid sinus. The

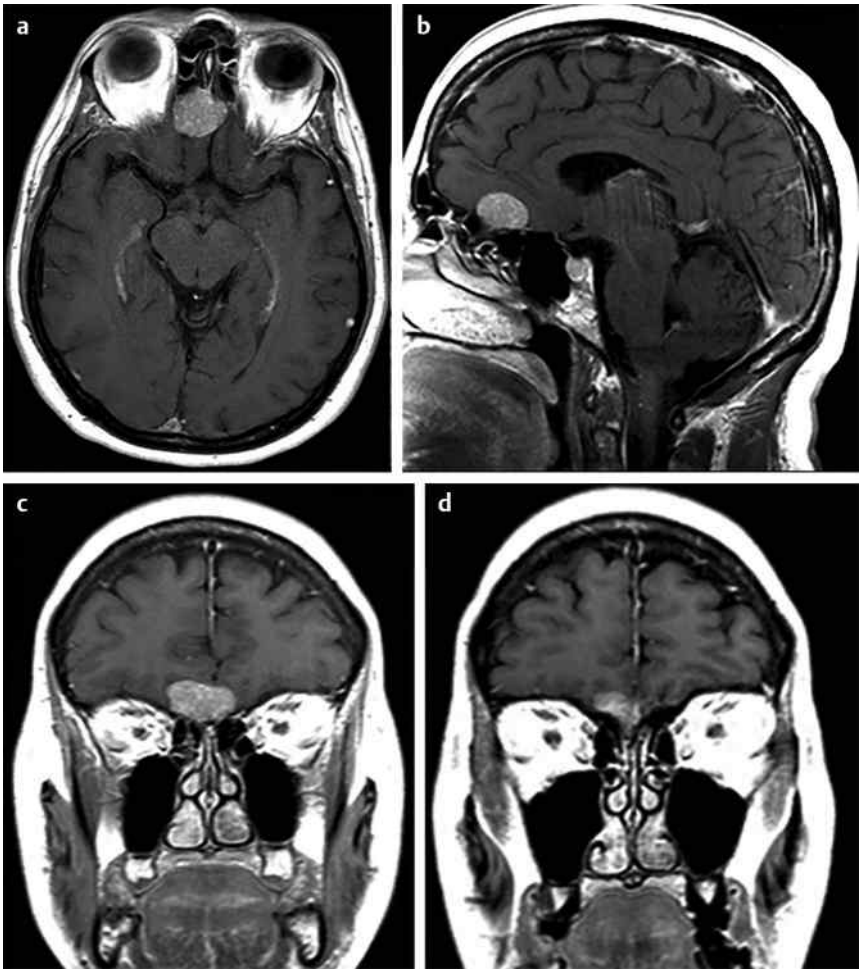

Fig. 2.1 Preoperative MRI. (a) Axial and **(b)** sagittal images show the 24 × 26 mm olfactory groove meningioma. **(c, d)** Coronal images show the right-sided origin of the tumor base.

differential diagnosis includes meningioma, lymphoma, granulomatous lesions, esthesioneuroblastoma, or dural-based metastasis. Dural metastases resembling meningiomas are rare, but most notably arise from prostate, breast, and kidney primary cancers. Based on the MRI characteristics, the slow growth rate of this tumor over 2 years, and the partial loss of olfaction, the most likely diagnosis in this case was an olfactory groove meningioma (OGM).

OGM accounts for about 8 to 13% of all intracranial meningiomas. They originate near the anterior cranial base, most commonly at the cribriform plate of the ethmoid bone, planum sphenoidale, and the frontosphenoidal suture. They can be in the midline, or eccentric to one side. OGMs should be differentiated from tuberculum sellae meningiomas, which are located more posteriorly over the tuberculum sellae, and present with visual disturbances due to early displacement of the optic apparatus. Sagittal cuts on MRI can be used to make this distinction. Blood supply of OGMs comes mostly from the ethmoidal arteries, and since these anastomose with ophthalmic arteries, embolization is not performed for tumors in this location because of the risk of blindness from accidental penetration of embolic material into the ophthalmic arteries. The tumor in this case originated unilaterally, from the right olfactory groove within the cribriform plate as seen on the coronal MRI.

OGMs often grow to a large size before causing symptoms, and in such cases, require surgery due to mass effect. For small, asymptomatic tumors that are found incidentally on imaging, these can often be managed with observation and repeat imaging. Surgery is recommended for documented growth in small lesions. Stereotactic radiosurgery (SRS) is a treatment option; however, the impact on the olfactory function is unpredictable. SRS is often reserved for older patients (> 80 years old) or those who cannot tolerate surgery.

Once surgery is selected as the treatment of choice, the goal for OGMs is usually to achieve a Simpson grade 1 resection to minimize the risk of future tumor recurrence. This translates to radical tumor removal along with underlying dural attachments and tumor infiltrated bone. In cases of OGMs less than 3 cm with preoperative hyposmia or normosmia, contralateral olfaction preservation is possible.

In the case described, the decision for surgery was made based on the documented radiographic growth over time, as well as the patient's symptoms of headaches and partial loss of olfaction. The partial or unilateral loss of olfaction indicated that the ipsilateral olfactory nerve has already been infiltrated by the tumor, but that the contralateral nerve was still functioning.

It is the senior author's (SY) practice to evaluate the bilateral olfactory function in OGM patients pre- and postoperatively with the University of Pennsylvania Smell Identification Test (UPSIT). This is a highly reliable, "scratch and sniff" test to determine function of each olfactory nerve. In this case, the patient scored 19/40 on the contralateral nostril, which was interpreted as microsmia. Objective and subjective documentation of partially intact olfaction in this patient dictate that surgical goals should include preservation of the contralateral olfactory apparatus in addition to Simpson grade 1 tumor resection.

■ Anatomical and Therapeutic Considerations

Options for Approach

OGMs may be resected via endoscopic or open craniotomy procedures. Common open approaches include the bifrontal and anterolateral craniotomies. The advantages of the bifrontal craniotomy are that it allows for full, bilateral access to the tumor through either an interhemispheric or subfrontal corridor. This provides excellent exposure, but in order to elevate and separate the frontal lobes, the superior sagittal sinus (SSS) has to be ligated to cut the falx. This carries a small risk of cerebral edema and venous infarction. Other disadvantages of this approach include opening both frontal sinuses, bifrontal manipulation and retraction, and late identification of the anterior cerebral arteries in large tumors.

Alternatively, the anterolateral approaches provide tumor access via a unilateral subfrontal or transsylvian route. Violation of both frontal sinuses, and manipulation of the contralateral frontal lobe and SSS can be avoided with these approaches. In addition, early identification and protection of the anterior cerebral arteries is a major advantage of using the anterolateral corridor. The drawbacks of attacking the tumor from the side, even with orbital osteotomies to increase the working space, include limited access to the base of the tumor within the cribriform plate and ethmoid sinuses, making a Simpson grade 1 resection challenging.

Regardless of the open cranial approach chosen, they all share the disadvantages of brain retraction and require tumor debulking prior to coagulating the arterial blood supply at the skull base for tumor devascularization. This can prolong surgical time, requires manipulation of neurovascular structures, and may result in frontal lobe edema, venous infarction, subdural hygromas, or hematomas.

With improvement of equipment and skills, endoscopic endonasal approaches (EEAs) are increasingly being considered as equal if not better approaches for OGMs compared to open approaches. Advantages of this approach include early control of tumor blood supply from anterior and posterior ethmoidal arteries, and definite resection of abnormal bone and dura at the base of the tumor. It is also superior to the transcranial routes for decompressing the optic nerves for tumors that involve the medial optic canal. In such cases, the bony exposure is extended to include extradural drilling of the medial wall of the involved optic canal and incision of the medial optic sheath. Tumor resection is performed without frontal lobe retraction and with minimal neurovascular manipulation, so not surprisingly, the endoscopic approach results in less postoperative frontal lobe edema. Disadvantages include a higher rate of cerebrospinal fluid (CSF) leak requiring a multilayer closure of the anterior skull base. It is also limited in application to small- and moderate-sized tumors.

Olfaction is a big concern for the EEA. The nasal cavity contains multiple sites of epithelium with olfactory afferents, which also must be preserved for olfactory function. These areas include the epithelial coverings of the cribriform plate, superior posterior nasal septum, superior turbinate, and medial aspect of the middle turbinate. The traditional binostril endonasal approach necessitates the

removal of both cribriform plates and often damages the occult olfactory epithelium high in the nasal cavity. The result of all this leads to anosmia.

To solve this problem, the unilateral transcribriform approach with septal transposition has been developed. In this approach, the olfactory epithelium is only disrupted on one side and thus olfaction can be preserved (**Fig. 2.2**). However, optimal patient selection is the key to success, and OGMs with bilateral cribriform origin would always necessitate opening both cribriform plates, thus disrupting bilateral olfaction (**Fig. 2.3**). For a patient to be considered for olfaction preservation, the following criteria must be met: the tumor must be less than 3 cm, have a *unilateral* origin within the cribriform plate, and the patient must have intact contralateral olfactory function. For patients fitting these criteria, OGM resection should be performed via an ipsilateral transcribriform approach, with a septal transposition to preserve the contralateral nasal epithelium.

Approach of Choice

Our patient, while needing a Simpson grade I resection, fits the criteria for consideration of olfaction preservation; her tumor is small and it is only attached to the right cribriform plate. Most importantly, her olfaction remained partially intact. For these reasons, the unilateral transcribriform approach with septal transposition was chosen as the approach of choice.

Questions

1. What extradural and intradural maneuvers must be carefully performed to preserve olfaction in this patient?
2. Is there a role for lumbar drainage in preventing a CSF leak in this patient?

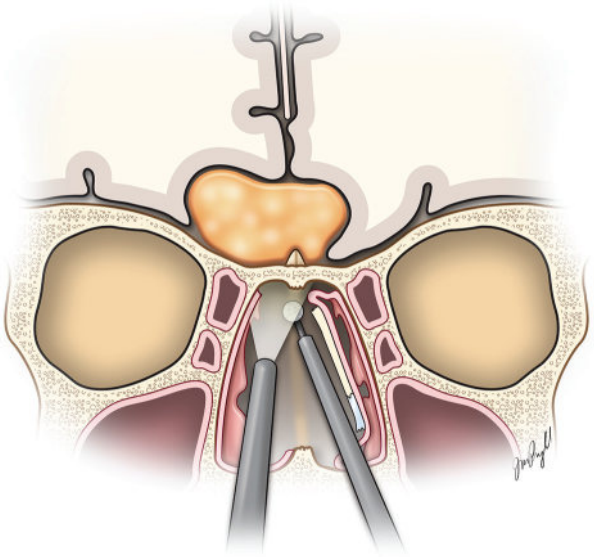

Fig. 2.2 Illustration of the endoscopic unilateral transcribriform approach. By preserving the mucosa on the left side during the extracranial phase, and protecting the left olfactory bulb and nerve during the intracranial dissection, olfaction can be maintained after surgery.

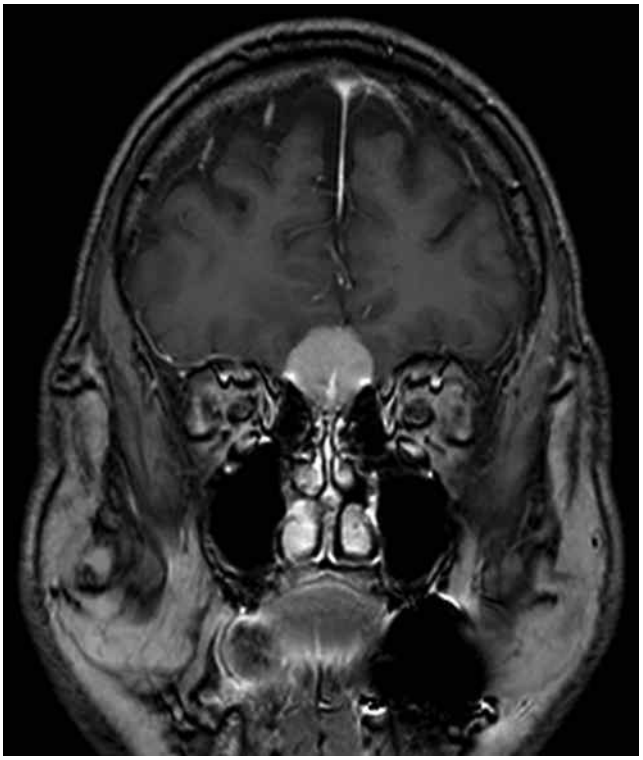

Fig. 2.3 Coronal MRI of a different patient. This shows a *bilateral* origin/base of the OGM with extension of the attachment into bilateral ethmoid sinuses.

■ Description of the Technique

The patient was positioned supine on the operating table with the head slightly extended in a head rest. Neuronavigation was employed using the contrast-enhanced, preoperative MRI with 1-mm axial sections (Medtronic Electromagnetic Fusion ENT Surgical Navigation, Louisville, CO). The nasal cavity was prepped by inserting pledgets soaked in oxymetazoline bilaterally for 5 minutes (see **The Three Approach Elements**).

A nasoseptal flap was mobilized on the right side and tucked in the nasopharynx for later reconstruction of the skull base defect. The septal transposition technique was utilized to facilitate binostril four-hand surgical access while preserving olfactory epitheliumon the contralateral side. A left-sided hemitransfixion incision was used. The olfactory epithelium was carefully elevated off the septum, including the epithelium from the perpendicular plate of the ethmoid bone and from the olfactory fossa. This preserves the olfactory neuronal inputs. The central and superior portion of the septal cartilage and bone were resected, allowing binostril access to the right cribriform plate. The right middle turbinate was resected and ethmoid air cells were opened in order to expand the working window (see **Operative Setup**).

The Three Approach Elements

Corridor: endonasal

Craniotomy: N/A

Modifier: septal transposition

Operative Setup

Position: supine

Incision: endonasal approach with planned nasoseptal flap

Bone Opening: unilateral right cribriform plate

Durotomy: dural base of the tumor

The cribriform plate on the right was resected using the Sonopet (Stryker, Kalamazoo, MI). Bone was removed beginning medially and working laterally to expose the dura underlying the lateral extent of the tumor. An angled tip was used to trim the medial edge of the cribriform plate and avoid the violation of the contralateral olfactory bulb/tract.

The dural base of the tumor and area surrounding the tumor was coagulated along with the anterior ethmoidal artery, which was the main blood supply to OGMs. This achieved early devascularization of the tumor, which was one of the benefits of this approach as compared to the open approaches. The dura was then sharply opened with a dural knife circumferentially around the tumor.

Once the dura was opened, the tumor was debulked centrally using the Sonopet. With sufficient internal debulking, the tumor capsule was then mobilized from the surrounding fronto-orbital cortex using a bimanual dissection technique. Thin cottonoids were placed into this dissection plane (**Fig. 2.4a**).

The dural base of the tumor extending past midline to the contralateral side was sharply incised, exposing the contralateral olfactory tract and orbitofrontal artery (**Fig. 2.4b**). This olfactory tract and its surrounding arachnoid plane must be carefully preserved to maintain its microvascular blood supply (**Fig. 2.4c**). The tumor capsule plane was carried over to the right side, and using sharp dissection, it was removed from the fronto-orbital cortex (**Fig. 2.4d**).

Once hemostasis was achieved, a multilayer closure was performed. The senior author's preference for this is a gasket seal closure with a PDS plate (KLS Martin, Jacksonville, FL) and DuraGen (Integra LifeSciences Corporation, Plainsboro, NJ). The nasoseptal flap that was created in the beginning of the procedure on the ipsilateral side was then mobilized and placed onto the skull base defect. The dislocated nasal septum was realigned in normal anatomical position, and the left side hemitransfixion incision was closed.

An edited version of the video from this operation accompanies this text (**Video 2.1**).

Surgical Pearls

1. As surgeons, we must unlearn what has become reflexive in our minds and decouple the link between surgery on OGMs and anosmia. Assessment of olfaction with the UPSIT provides objective measurement of the sense of smell, and in selective patients with small OGM, preservation of function must be considered in addition to a Simpson grade I resection. A unilateral approach with septal mobilization to preserve the contralateral epithelium, and a transcribriform access using only the affected side can in fact preserve olfaction.
2. Once intradural, the mission to preserve all intact neurological function, not just olfaction, requires meticulous bimanual dissection of neurovascular structures, respecting the arachnoidal planes. This minimizes the risk to the orbitofrontal arteries, cortices, and the contralateral olfactory bulb and tract.
3. Multilayered reconstruction of the skull base defect is critical in avoiding postoperative CSF leak.

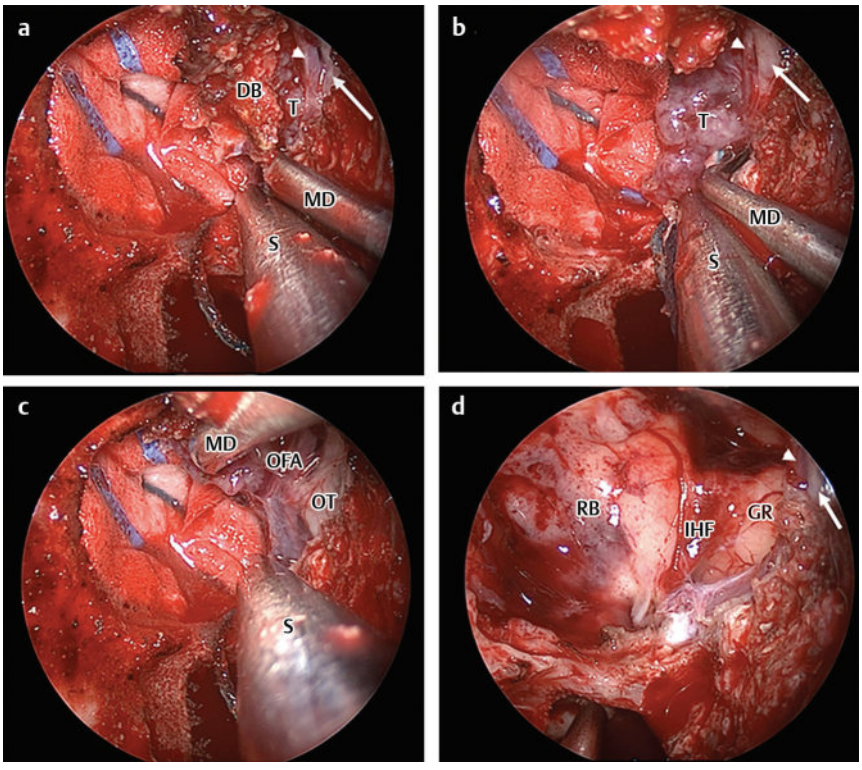

Fig. 2.4 Intraoperative images. (a) Right nostril approach showing a dural base resection. **(b)** Tumor dissection from contralateral olfactory tract. **(c)** Intact arachnoid plane around olfactory tract. **(d)** Postresection, intact neurovascular bundle. DB, dural base; GR, contralateral gyrus rectus; IHF, interhemispheric fissure; MD, microdissector; OFA, orbitofrontal artery; RB, resection bed; S, suction tip; T, tumor; arrow, contralateral olfactory tract (OT); arrowhead, contralateral orbitofrontal artery (OFA).

■ Aftercare

Our patient had an uncomplicated postoperative course, and the MRI after surgery showed no residual tumor as a Simpson grade I resection was achieved (**Fig. 2.5**). Six months after surgery, an outpatient endoscopy was performed. The left nasal cavity had no evidence of disruption of mucosal lining. On the right, the nasoseptal flap was at the appropriate stage of healing (**Fig. 2.6**).

■ Possible Complications and Associated Management

Potential complications of this procedure include those related to any EEA for an intradural lesion, particularly CSF leak. With multilayered skull base reconstruction and a vascularized mucosal flap, a lumbar drain is not necessary. Postoperatively, the patient should be monitored in the intensive care unit (ICU) with frequent neurological checks and intravenous Decadron at a standard dosage of 4 mg every 6 hours for 24 to 48 hours, depending on the amount of preexisting cerebral edema.

When examining a patient after this procedure with a nasoseptal flap, it is preferable not to attempt to induce CSF leak with provocative testing (i.e., reservoir test). This can sometimes cause the nasoseptal flap and other closure layers to become dislodged, causing the very CSF leak that one worked so hard to prevent. If postoperative CSF rhinorrhea is evident despite all the preventive measures, the patient should be taken back to the OR for inspection of closure and repositioning of the nasoseptal flap. Lumbar drain or external ventricular drain should not be placed, as this can cause pneumocephalus.

A common practice with transdural endoscopic procedures is to use broad-spectrum, triple antibiotics in the postoperative period for 48 to 72 hours. This is based on the hypothesis that meningitis most likely occurs after intradural surgeries, nasal packing, and preexisting sinus disease.

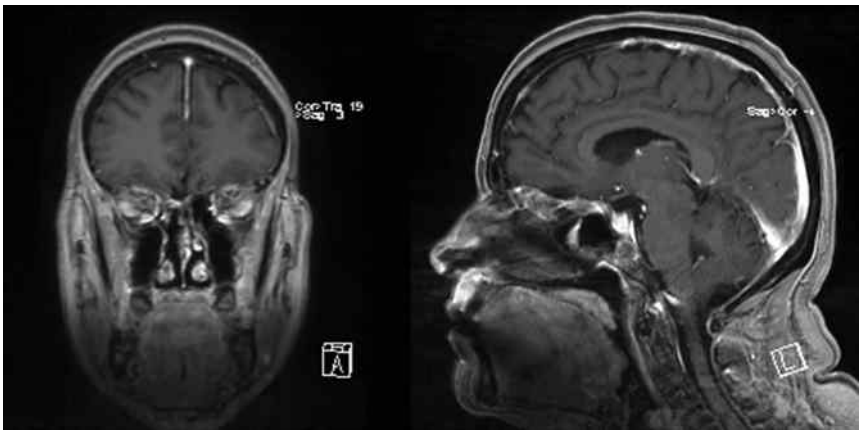

Fig. 2.5 Postoperative MRI. Coronal and sagittal images showing Simpson grade I resection of the tumor.

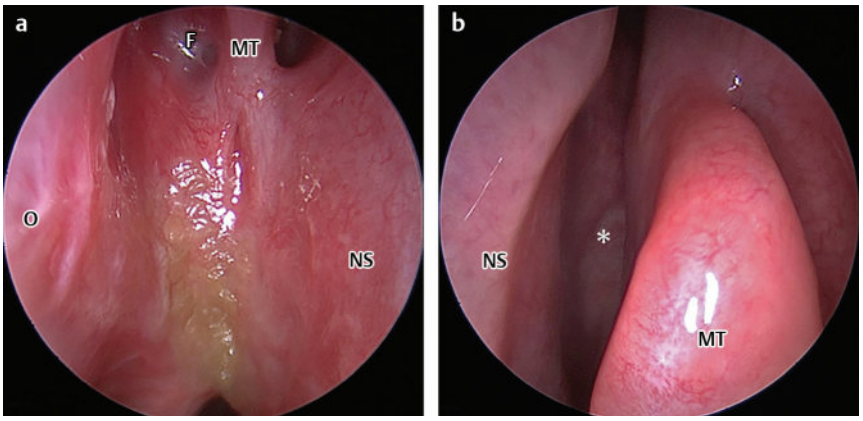

Fig. 2.6 Six-month postoperative endoscopic images. (a) Right nasal cavity with a 30-degree endoscope demonstrating scarring at the healed surgical site. **(b)** Normal appearance of left nasal cavity including olfactory cleft (*). F, frontal sinus; MT, partially resected middle turbinate; NS, nasal septum; O, orbit.

Perspective

Michael C. Huang

■ Introduction

OGMs constitute only approximately 10% of intracranial meningiomas and because of their rarity and insidious clinical course, they are often discovered only after significant growth. In fact, the mean tumor size at the time of surgery ranges from 4.46 to 5.4 cm. Not surprisingly, headaches, visual loss, and mental status changes are the most commonly associated symptoms, but included among these is anosmia. While surgical resection of OGM can improve preoperative visual, cognitive, and psychological dysfunctions, surgery does not restore lost olfaction and rarely improves dysfunctional olfaction. Even when the anatomical continuity of the olfactory tracts is maintained during the operation, functional olfaction is not guaranteed.

In the previous section, the operation to preserve intact olfaction was thoroughly discussed, but how does one make surgical decisions when an OGM presents in the more common scenario, when olfaction is lost? A multitude of surgical approaches have been described for the surgical management of OGM with the goal of complete tumor resection along with associated dura and infiltrated hyperostotic bone. These approaches include bifrontal and unilateral frontal craniotomies with subfrontal approach, pterional craniotomies with transsylvian approach, and transbasal approaches. More recently, several authors have reported on the application of EEA for these tumors. As always, appropriate approach selection, tailored to tumor size, is critical for the success of surgery and limiting morbidity.

■ Case Presentation

A 43-year-old man presented to neurosurgery clinic with 8-month history of progressive visual loss. The patient and his wife have also noted cognitive slowing and memory difficulties over the same period. On examination, he was found to have an afferent pupillary defect with only light perception in the left eye and significantly decreased vision in the right eye. An MRI of the brain revealed a large 7.5 × 7.5 × 6.5 cm avidly enhancing extra-axial tumor arising from the anterior skull base consistent with an OGM (**Fig. 2.7**).

■ Anatomical and Therapeutic Considerations

The MRI demonstrated that the tumor occupied the entire anterior skull base, extending from the inner table of the frontal sinus to the tuberculum sellae (**Fig. 2.7**). The optic chiasm has been pushed inferiorly and posteriorly. This feature of OGM distinguishes it from other meningiomas of the anterior skull base, such as tuberculum sellae meningiomas, which usually elevates the optic chiasm. The tumor extended superiorly from the midline to displace symmetrically both frontal lobes (**Fig. 2.8**). There were numerous flow voids, especially at the level of the cribriform plate, and a cerebral angiogram confirmed the highly vascular nature of the tumor with primary arterial supply via bilateral anterior and posterior ethmoidal arteries, and recurrent meningeal arteries (**Fig. 2.9**). The anterior cerebral arteries were displaced superiorly. A computed tomography scan showed hyperostosis along the cribriform plate and crista galli. There was no tumor extension inferiorly into the ethmoidal sinuses.

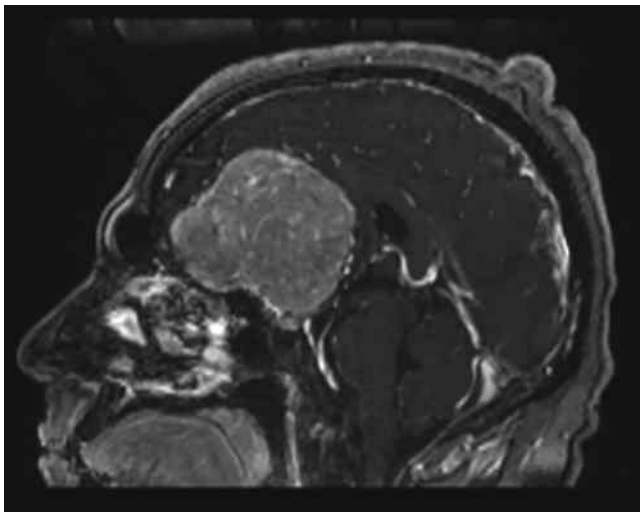

Fig. 2.7 Preoperative MRI. This showed a 7.5 × 7.5 × 6.5 cm avidly enhancing extra-axial tumor arising from the anterior skull base consistent with an olfactory groove meningioma. The tumor abutted the inner table of the frontal sinus anteriorly and extended to the dorsum sellae posteriorly. There was no invasion of the ethmoid sinus.

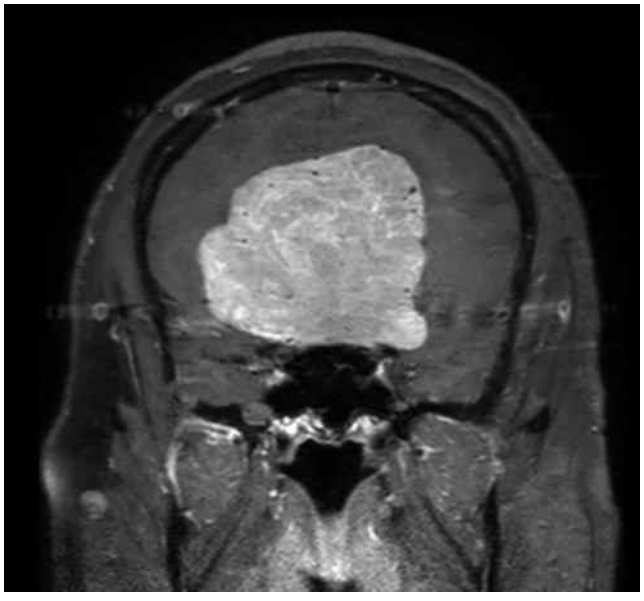

Fig. 2.8 Preoperative MRI. This showed the tumor projecting superiorly from midline and extending laterally in a fairly symmetrical fashion.

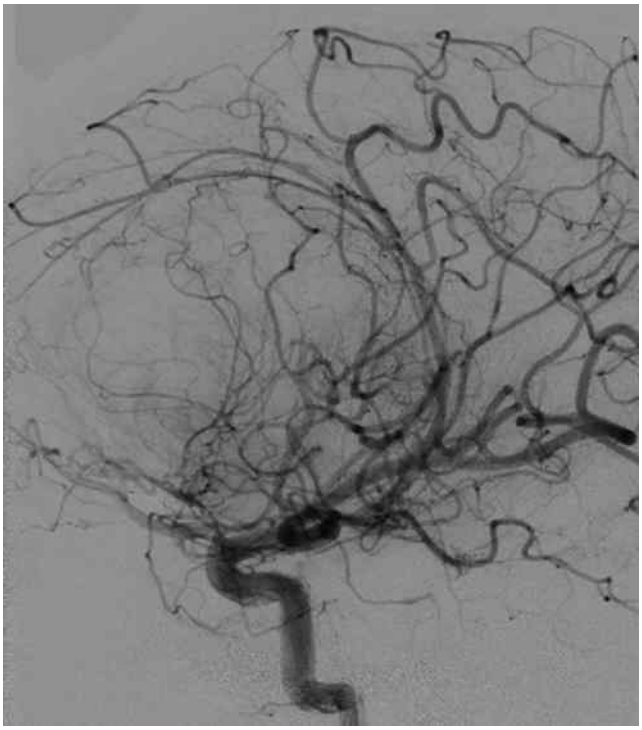

Fig. 2.9 Preoperative cerebral angiogram. The right internal carotid artery injection demonstrated a large vascular lesion at the anterior skull base. The feeding ethmoidal arteries were enlarged. The anterior cerebral arteries were displaced posteriorly and superiorly.

Options for Approach

Given the size of the tumor and the patient's progressive visual loss, surgical resection was indicated. Traditionally, an anterior approach via a bifrontal craniotomy is the recommended surgical strategy for addressing large OGMs. A bifrontal craniotomy offers panoramic access to nearly all sides of the tumor and provides good visualization of the anterior cranial floor, allowing for early disruption of the ethmoidal arteries, complete resection of hyperostotic bone, and straightforward cranial reconstruction. However, when compared to other approaches to OGM, bifrontal craniotomies are associated with increased risk of life-threatening complications, such as malignant cerebral edema and postoperative hemorrhage. This may be the result of transection of the anterior third of the SSS, which is commonly performed during the bifrontal approach and considered harmless. Furthermore, even when the anterior cut of the frontal craniotomy is taken as far inferiorly as possible above the orbital rim, retraction of the frontal lobes may still be necessary in order to work over the residual bony lip. Frontal lobe changes, including porencephalic cave formations, are more likely to be seen on postoperative imaging following a bifrontal approach. Other disadvantages of bifrontal craniotomy include late visualization of the optic chiasm and anterior cerebral arteries, violation of the frontal sinuses with risk of CSF leak, and blind reach of tumor extending below the tuberculum sellae.

While initially utilized for small- to medium-sized tumors, several authors have reported high rates of Simpson grades I and II resection of large OGMs through a unilateral approach via pterional craniotomies. Approaching the tumor through the anterolateral corridor allows for early aspiration of CSF from the basal cisterns to promote cerebral relaxation, alleviating the need for frontal lobe retraction. The optic apparatus and the supraclinoid internal carotid artery are exposed at the start of tumor dissection and the tumor is devascularized by coagulating the basal attachment across the anterior skull base. Resection of the tumor from a unilateral approach obviates the need for transection of the SSS and requires manipulation of only the ipsilateral frontal lobe and olfactory tract. The challenges of the pterional approach include a relatively narrow working angle with partial obstruction from an elevated and rough orbital roof. The exposures to the contralateral side of the tumor and to the anterior cerebral arteries draped over the superior capsule of the tumor are potentially inadequate.

Both the bifrontal and pterional craniotomies can be expanded with the application of skull base techniques and modifications of the bone opening. The addition of basal osteotomies, such as unilateral or bilateral orbital osteotomies, provides more tangential trajectory to the cranial base, shortens the working distance to the tumor, widens the working angle, especially to the superior portion of the tumor, and minimizes the need for brain retraction. Extensive basal osteotomies and transfacial approaches can facilitate radical resection of tumor extending into the ethmoid sinuses and nasal cavity along with affected bone. These benefits must be balanced against increased surgical complexity and time, and higher risks of CSF leaks.

Recent advancements in endoscopic techniques have significantly expanded the surgical armamentarium of the cranial base surgeon. EEA to OGM resection offers wide illumination of the skull base with early access to vascular pedicles, and to tumor infiltrated bone and dura. With direct visualization of the neurovascular structures from below, the tumor can be safely resected with minimal brain manipulation. However, large tumors, especially those with considerable lateral extensions, remain challenging for endoscopic techniques. Endoscopic surgeries for large OGM are associated with high rates of staged surgeries and lower rates of gross total resection compared to open approaches. Furthermore, even with the use of nasal septal flaps, the rates of CSF leak following EEA remain concerning.

Approach of Choice

In this case, given the large size of the tumor with symmetric extensive lateral extension over the bilateral orbital roofs, significant superior extension with the anterior cerebral arteries displaced over the tumor's superior pole, the decision was for a transbasal approach with bifrontal craniotomy and bilateral orbital osteotomies. In order to avoid potentially disfiguring frontal osteotomy above the orbital rims and to simplify reconstruction, a one-piece transbasal approach was planned (**Fig. 2.10**).

■ Description of the Technique

A lumbar drain was placed following the induction of general anesthesia. The patient was positioned supine with his neck gently flexed. A bicoronal scalp incision was made and the pericranium was harvested and protected. The supraorbital neurovascular bundles were mobilized from their foramens, and the periorbita was dissected free from the orbital roof bilaterally. Bilateral MacCarty burr holes were placed along with two burr holes straddling the midline posteriorly.

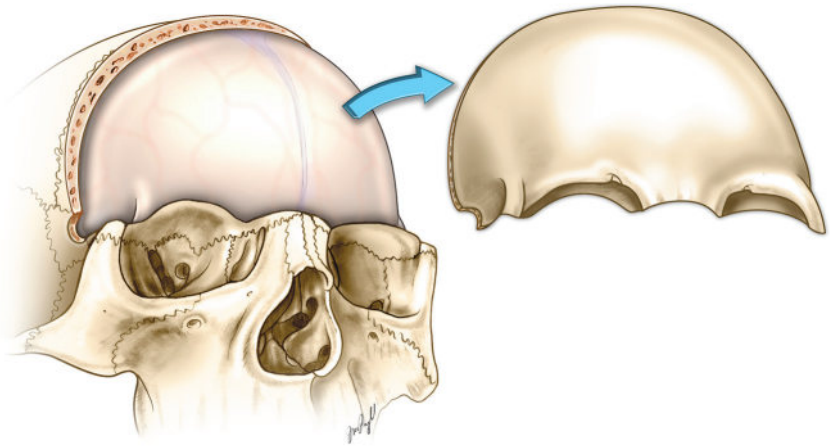

Fig. 2.10 Illustration of the one-piece transbasal craniotomy.

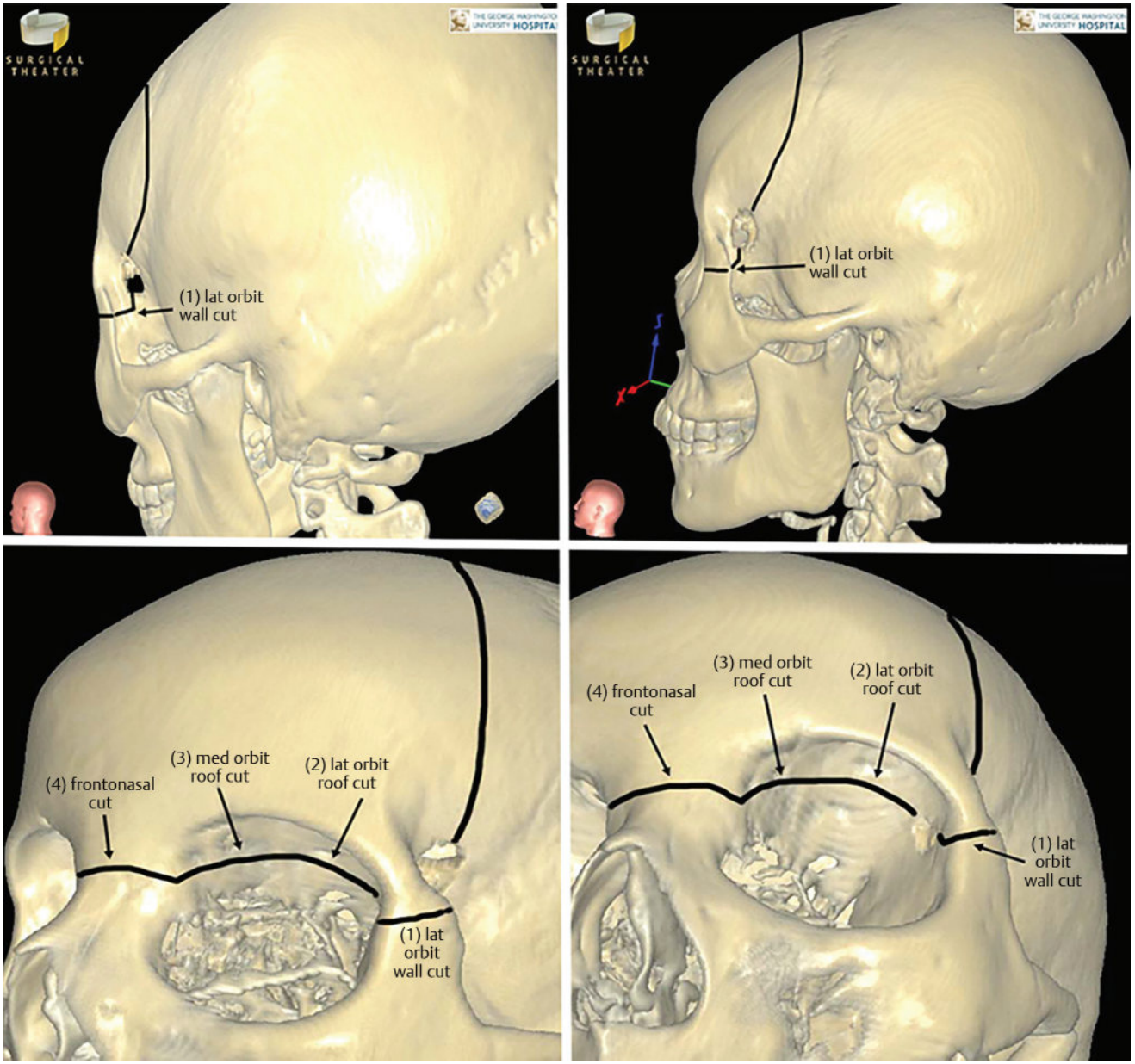
SURGICAL THEATER
(1) lat orbit wall cut
SURGICAL THEATER
(1) lat orbit wall cut
(4) frontonasal cut
(3) med orbit roof cut
(2) lat orbit roof cut
(1) lat orbit wall cut
(4) frontonasal cut
(3) med orbit roof cut
(2) lat orbit roof cut
(1) lat orbit wall cut

Fig. 2.11 Virtual reality-generated images demonstrating the orbital cuts for the one-piece transbasal approach. Posterolateral, lateral, anterolateral, and anteromedial views showing the cuts from the MacCarty burr hole. (1) Lateral orbital wall, (2) lateral orbital roof, (3) medial orbital roof, and (4) frontonasal cuts are detailed in the text. (Figure by Dr. Walter Jean.)

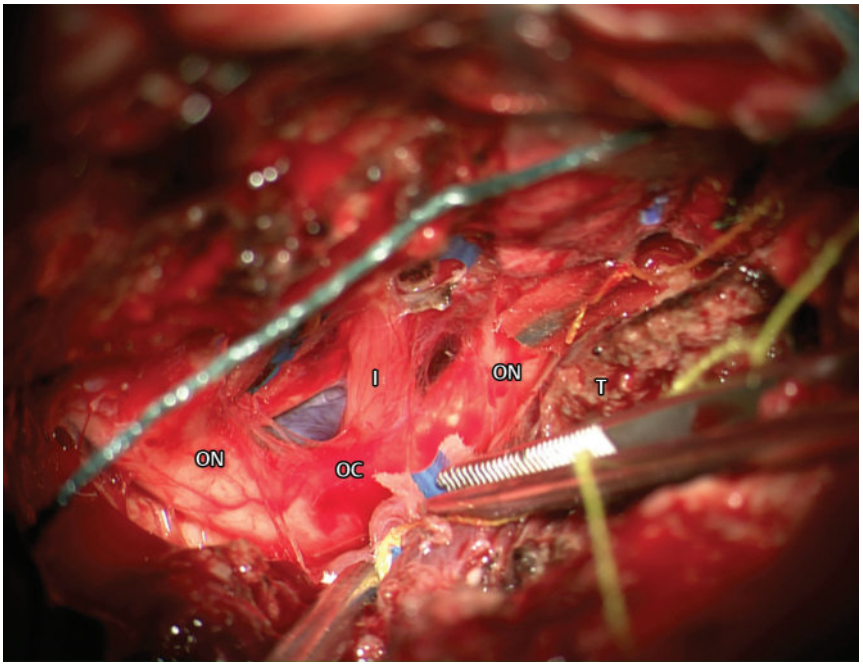

Fig. 2.12 Intraoperative image. This shows the dissection of the tumor off of optic apparatus. I, infundibulum; OC, optic chiasm; ON, optic nerve; T, tumor.

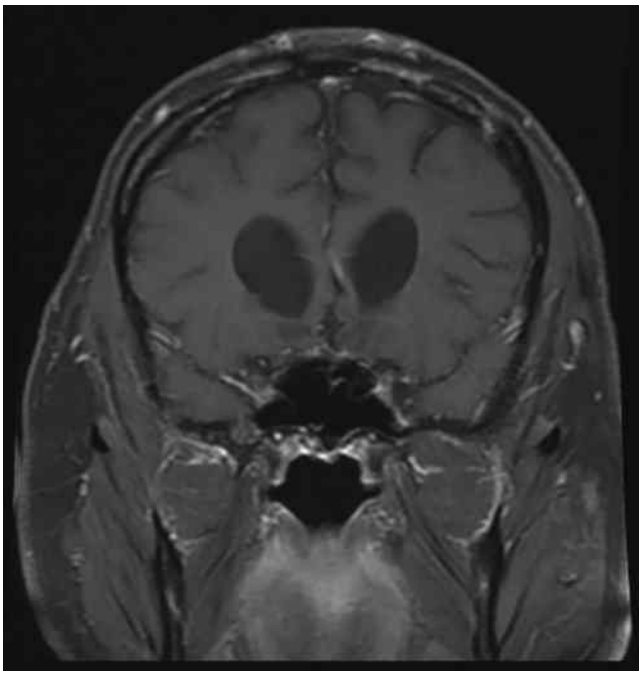

Fig. 2.13 Postoperative MRI. This image at 30 months demonstrates no recurrence of tumor.

Once the SSS was dissected free, the four burr holes were connected. Using a craniotome, bilateral orbital rim cuts were made starting from the orbital portion of the MacCarty burr hole down to the level of the frontozygomatic suture (step 1, **Fig. 2.11**). With the periorbita protected and the globes gently depressed, intraorbital cuts were made medially across the posterolateral orbital roof starting from the MacCarty burr holes using a small osteotome (step 2, **Fig. 2.11**). The same osteotomes were used for the medial orbital roof cuts, done from within the orbit, with the periorbita protected, aiming superiorly and medially (step 3, **Fig. 2.11**). These cuts were made at the level of the frontonasal suture. The final cut was made across the frontonasal suture, above the medial canthal ligaments, using a side-cutting drill with a straight guard directed 30 degrees superiorly, connecting the bilateral intraorbital cuts (step 4, **Fig. 2.11**). The frontal bone along with the orbital bandeau was elevated as a single piece.

The crista galli was identified and removed. Intraorbital dissection was performed and the ethmoidal arteries were identified and divided. With the assistance of CSF drainage via the lumbar drain for brain relaxation, the frontal dura was elevated from the anterior skull base, as additional perforators were coagulated. The dura was opened over the anterior frontal poles and the SSS was ligated.

The bilateral frontal lobes were displaced superiorly and posteriorly by the tumor, but there was a preserved arachnoid plane between the tumor and the frontal poles. The tumor was entered along its skull base attachment. Despite early transection of the ethmoidal feeders, the tumor remained quite vascular. The tumor was internally debulked and the tumor capsule progressively mobilized. The optic nerves were flattened and thinned by the tumor, but the arachnoid plane was intact (**Fig. 2.12**). The anterior communicating artery complex, distal anterior cerebral arteries and perforators were carefully protected. Tumor-feeding vessels from the anterior cerebral arteries were traced to their termination in the tumor before they were divided. This reduced the bleeding from the tumor significantly, and the remaining tumor was removed without further difficulties. The affected dura at the anterior skull base was resected, and the underlying hyperostotic bone was drilled flat with a diamond drill bit.

The dural defect at the skull base was repaired with a patch, and the dura was closed in a watertight fashion. The frontonasal ducts were plugged with muscle and the previously harvested pericranial flap was reflected down over the skull base and tacked to the basal frontal dura. The frontal sinus on the bone flap was cranialized and the mucosa was removed. The bone flap was secured back in place with care not to compress the pericranial flap.

Aftercare

The patient was monitored in the ICU postoperatively. His lumbar drain was kept clamped and then removed when there was no evidence of CSF leak. A postoperative MRI showed total resection and follow-up imaging at 30 months showed no recurrence (**Fig. 2.13**). Patient's visual acuity improved to 20/20 OD and 20/30 OS at 6 months postoperatively. His cognitive impairments had completely resolved and he has returned to work full time.

Commentary

The surgical treatment of OGM has evolved significantly since the first report of successful resection by Francis Durante in 1887. Advancements in microsurgery, cranial base approaches, and endoscopic techniques have greatly expanded the surgical options for the modern neurosurgeon. However, the treatment of OGM remains challenging since the majority of patients present when the tumor is huge, which increases the likelihood of neurovascular involvement and operative complications. Complete surgical resection has been shown to be critical for prevention of recurrence. At present, no single best surgical approach exists and the various surgical options should be viewed as complimentary rather than mutually exclusive. One such option is to perform surgery in staged fashion, with the first stage done endoscopically specifically to eliminate the tumor feeders through the cribriform plate. Internal debulking would also be accomplished in this stage, and the usual concern for postoperative hemorrhage is mitigated by the devascularization step performed before entering the tumor. A craniotomy would be planned as a second stage, with the intent of being "minimally invasive," made feasible by the devascularization accomplished in the first stage. Theoretically then, instead of a large transbasal approach, our patient might be a candidate for a "keyhole" operation in this multidirectional, staged scheme.

Recommended Reading

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3 Nasopharynx and Pterygopalatine Fossa

Lilun Li and Ameet Singh

Keywords: juvenile nasopharyngeal angiofibroma, endoscope, pterygopalatine fossa, infratemporal fossa, Caldwell–Luc maxillotomy

■ Case Presentation

A 26-year-old man presented with a 1-month history of nasal obstruction and left-sided epistaxis. The patient had no prior history of nasal trauma or sinusitis. Nasal saline, steroid sprays, and oral antihistamines did not relieve his obstructive symptoms. On endoscopic examination, the patient was found to have a fibrous-appearing mass filling the left nasal vault. A maxillofacial computed tomography (CT) scan (**Fig. 3.1**) revealed a large mass centered in the left nasal cavity extending to the ipsilateral paranasal sinuses, and central skull base. Heterogeneous uptake of gadolinium on magnetic resonance imaging (MRI) suggested a hypervascular mass (**Fig. 3.2d–f**).

Questions

1. Given the differential diagnosis in this 26-year-old man, is a biopsy indicated for this mass?
2. What characteristics on endoscopic examination would help in making the diagnosis?
3. Where did this mass originated from and what spaces did it spread into?

■ Diagnosis and Assessment

The differential diagnosis for a unilateral sinonasal mass in a young adult causing nasal obstruction and epistaxis is wide. The primary differential includes inverted papilloma, juvenile nasopharyngeal angiofibroma (JNA), antrochoanal polyp, hemangiopericytoma, and lymphoma. The patient's associated epistaxis, and imaging findings of a locally invasive and hypervascular mass, makes JNA the most likely diagnosis, although hemangiopericytoma cannot be ruled out.

JNAs are the most common benign tumor of the nasopharynx, and are almost exclusively seen in adolescent males. Although some controversy remains over the exact site of origin, these lesions tend to arise from the sphenopalatine foramen and spread anteriorly into the nasal cavity and posteriorly into the nasopharynx. They may spread laterally into pterygopalatine fossa (PPF), through the pterygomaxillary fissure into the infratemporal fossa (ITF), or posteriorly into the bony pterygoid plates. Less commonly, these locally aggressive tumors can extend, via the PPF and inferior orbital fissure into the orbit, or via the pterygoid plates into the middle cranial fossa. If the sphenoid sinus walls are eroded, the cavernous sinus and pituitary glands may become involved as well. Clinically, these tumors present with unilateral nasal obstruction and epistaxis, and a smooth, lobulated red or pale blue hypervascular lesion in the nasal vault is usually seen on endoscopic examination. CT scans often show bowing of the posterior maxillary sinus wall (Holman–Miller sign), widening of the PPF, bony erosion or remodeling of the skull base, and deviation of the nasal septal wall. On MRI, JNAs enhance brightly, often showing obvious flow voids on T2. Both characteristics are indicative of tumor hypervascularity.

Given its typical clinical presentation and radiographic findings, biopsies of JNA are controversial because of the risk of severe hemorrhage. The general consensus is that biopsy is unnecessary and too risky when suspicion is high, but some surgeons continue to advocate for pretreatment biopsy to avoid misdiagnosis of sinonasal malignancy in the setting of atypical clinical presentation. Though rare, solitary fibrous tumor/hemangiopericytoma (SFT) must be considered in the differential diagnosis due to a high distant metastasis rate of around 15%. These tumors are most commonly seen in middle-aged patients without significant gender discrepancy. One clue that would differentiate JNA from SFT is that the latter appears more fibrous than erythematous on endoscopy.

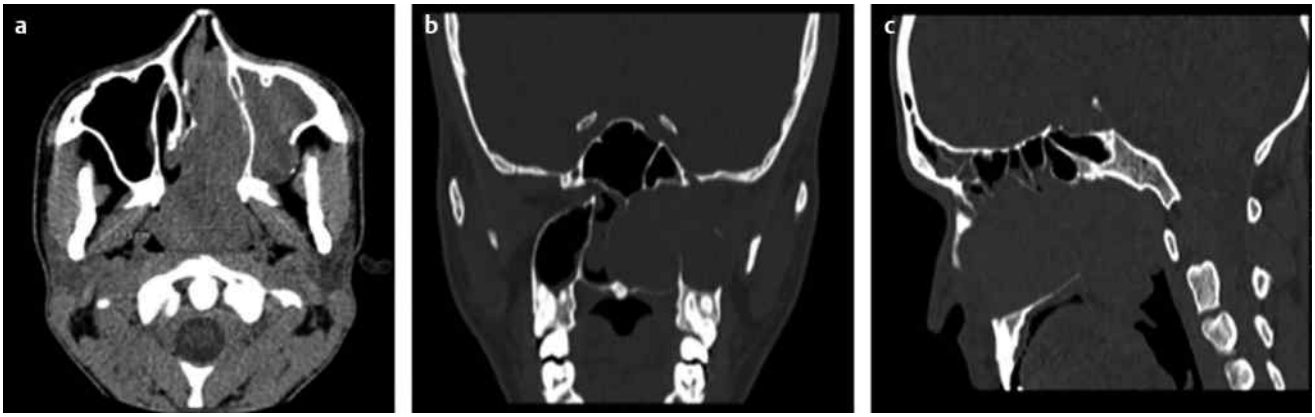

Fig. 3.1 Maxillofacial CT scan. (a) Axial soft tissue window CT showed a 7.9 × 3.7 cm mass extending from nasal cavity into the nasopharynx, pterygopalatine fossa, and left maxillary sinus. **(b)** Coronal bone window CT demonstrated erosion of the posterior and medial maxillary sinus wall as well as destruction of the left pterygoid plates. **(c)** Sagittal bone window CT showed no obvious invasion of the skull base.

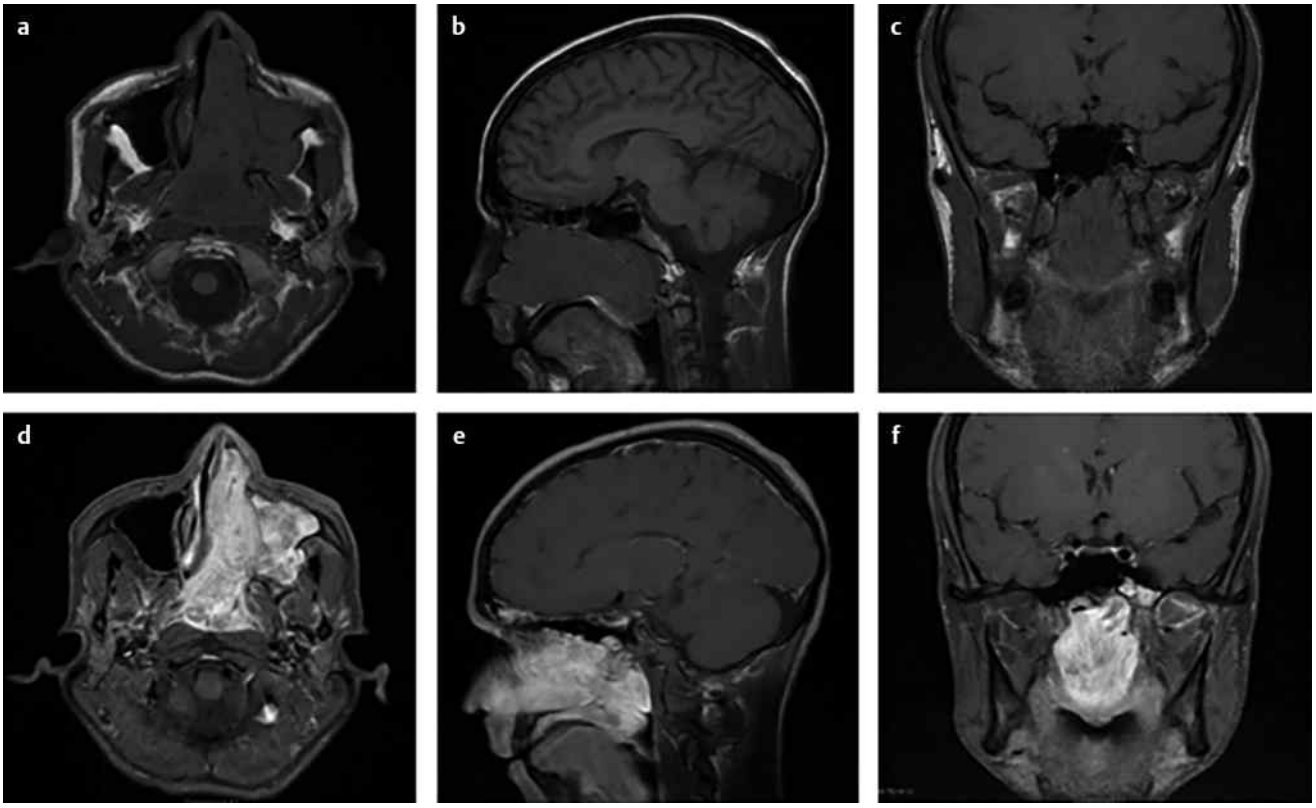

Fig. 3.2 Brain MRI with and without contrast. Precontrast T1-weighted MRI in **(a)** axial, **(b)** sagittal, and **(c)** coronal views showed lobulated mass measuring approximately 8.5 × 5.0 × 4.5 cm. The mass extended from the left nasal cavity and nasopharynx into the left posterior and medial maxillary sinus walls and left pterygoid plates, with extension into the left maxillary sinus, pterygopalatine fossa, and infratemporal fossa. Postcontrast T1-weighted MRI scans in the same views **(d–f)** demonstrated heterogeneous enhancement with irregular central hypointensity.

Our patient had an atypical presentation for JNA. First, the patient's age was above the upper limit of what is commonly seen with JNA, which has a reported average age of 15 years. Additionally, his initial endoscopic examination finding of a fibrous intranasal mass which did not fit the classic smooth, red or pale blue vascular appearance of JNA. As such, this patient underwent a nasal mass biopsy. He did not have any hemorrhagic complications, and the results showed vascular proliferation with thin-walled blood vessels, along with a fibrocollagenous stroma. No mitotic activity or cytologic atypia was seen. These findings were all suggestive of a diagnosis of JNA.

■ Anatomical and Therapeutic Considerations

Once diagnosed, JNAs are most often treated with surgical excision, with the goal of complete resection with minimal cosmetic deficit. With that goal in mind, the anatomy of the tumor was reviewed in detail. The tumor occupied the left nasal cavity and nasopharynx, and extended into the left maxillary sinus, PPF, and ITF (**Fig. 3.2**). On the CT bone windows (**Fig. 3.1b, c**), the mass remodeled and partially destroyed the left pterygoid plates, as well as the posterior and medial walls of the maxillary sinus. There was, however, no sign of erosion of the middle fossa floor.

The ITF is the anatomical space under the middle fossa floor (**Fig. 3.3**). Anteriorly, it is limited by the lateral part of the posterior wall of the maxillary sinus (PWMS), and its medial wall is formed by the lateral pterygoid plate. The ITF contains the pterygoid muscles, the maxillary artery, the pterygoid venous plexus, and branches of V3. Via the pterygomaxillary fissure, the ITF communicates with the PPF which is medial to it (**Fig. 3.4**).

The PPF is an inverted pyramid-shaped space behind the medial portion of the PWMS. It is bordered medially by the palatine bone and posteriorly by the pterygoid plate. Its apex, which is its inferior-most point, is the greater palatine canal. The contents of the PPF include the pterygopalatine ganglion, V2, the infraorbital and vidian nerves (**Fig. 3.5**).

Multiple staging systems have been developed based on the relationship between JNA and its surrounding anatomical structures. One of the early JNA staging systems proposed by Sessions was based on the degree of local tumor extension rather than JNA tumor size alone. A revised system, currently more popular, was proposed by Radkowski, and it distinguishes between isolated skull base erosion versus erosion with extensive intracranial and/or dural spread. Both JNA staging systems aimed to guide the amount of operative exposure necessary for full surgical resection, as well as the need for radiation therapy in unresectable tumors (**Table 3.1**).

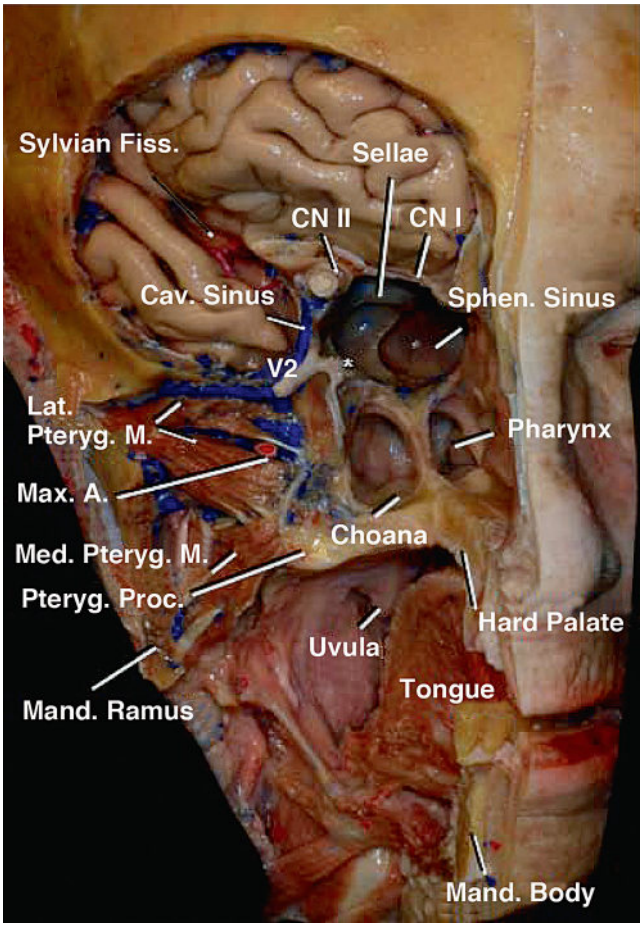

Fig. 3.3 Infratemporal and pterygopalatine fossa (IFT and PPF). Dissection of the posterior limit of the nasal cavity. The orbit has been resected up to the apex, and the walls of the maxillary sinus, the nasal septum, and part of the palate have been removed. The PPF, containing the pterygopalatine ganglion (*) and the IFT, with structures related to the posterior wall of the maxillary sinus, have been exposed. The sphenoid sinus has been opened. The anterior and middle fossae have been exposed, and part of the tongue, the ramus, and the body of the mandible have been removed. At the level of the choanae, the lateral relationship of the nasal cavity includes the PPF and IFT. A., artery; Cav., cavernous; CN, cranial nerve; Fiss., fissure; Lat., lateral; M., muscle; Mand., mandibular; Max., maxillary; Med., medial; Proc., process; Pteryg., pterygoid; Sphen., sphenoidal. (Reproduced from Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery. 1st ed. Thieme; 2011.)

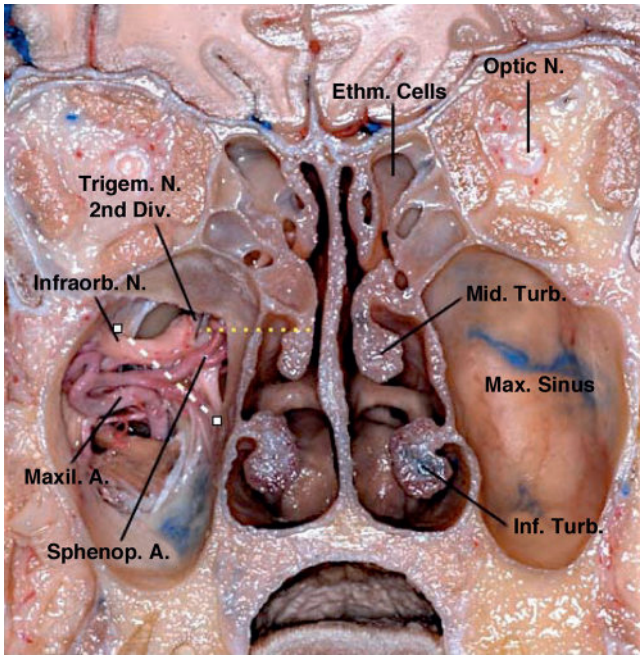

Fig. 3.4 Cranial base, coronal view. The posterior wall of the right maxillary sinus was removed, exposing the infratemporal fossa laterally, and the pterygopalatine fossa medially. The *white dashed line* is the demarcation of the two. The maxillary division of the trigeminal nerve (V2) is exiting the sphenoid bone through the foramen rotundum and running in the superior wall of the maxillary sinus as the infraorbital nerve. The maxillary artery loops in the infratemporal fossa, entering the pterygopalatine fossa and giving its terminal branches. The *yellow dotted line* demonstrates the relationship of the axis of the middle turbinate tail and the sphenopalatine foramen. A., artery; Div, division; Ethm., ethmoidal; Inf., inferior; Infraorb., infraorbital; Max./Maxil., maxillary; N., nerve; Sphenop., sphenopalatine; Trigem., trigeminal; Turb., turbinate.

As JNAs are believed to originate from the sphenopalatine foramen, all surgical approaches should be able to expose this anatomical landmark. Anterior and medial exposure into the nasopharynx and nasal cavity is also required, as these structures become involved early in the natural course of tumor spread. Lateral exposure is often necessary, as JNA extension into the PPF is reported between 26 and 100%, with further extension into the ITF reported between 20 and 54%. The incidence of intracranial involvement is reported at 10 to 20%, but extent of surgical exposure for these tumors depends on the level of invasion and involvement of intracranial structures. The significant morbidity associated with resection of tumors involving the internal carotid artery (ICA), optic nerve, or oculomotor nerves, makes partial resection with adjuvant radiotherapy worthy of consideration.

Options for Approach

Various open approaches have been developed to achieve adequate surgical exposure to facilitate total resection, while limiting intraoperative blood loss. A transpalatal approach can be used to resect tumors limited to the nasopharynx and nasal cavity, while a combined transpalatal and transantral approach can resect tumors that extend laterally into the PPF and/or maxillary sinuses. The combined transpalatal–transantral approach was found to effectively treat tumors of Radkowski's stage IB or less with minimal recurrence. Risks of transpalatal tumor resection include palate dehiscence and oronasal fistula. A combined transpalatal–transantral approach presents additional risks of facial deformity and paresthesia due to infraorbital nerve injury.

Lateral rhinotomy with extension to the upper lip mucosa came into favor for resection of more extensive tumors. This approach provides access to medial structures such as the nasal cavity and nasopharynx, as well as to lateral structures such as the maxillary sinus, PPF, pterygomaxillary fossa, and infratemporal region. Besides an unattractive nasal scar, other risks of the lateral rhinotomy include facial paresthesia in the infraorbital nerve distribution and lacrimal apparatus injury from the superior incision site.

A midfacial degloving technique was developed to address the cosmetic deformity problem presented by lateral rhinotomy; its intranasal and sublabial incisions provided excellent surgical exposure while eliminating external scarring. The

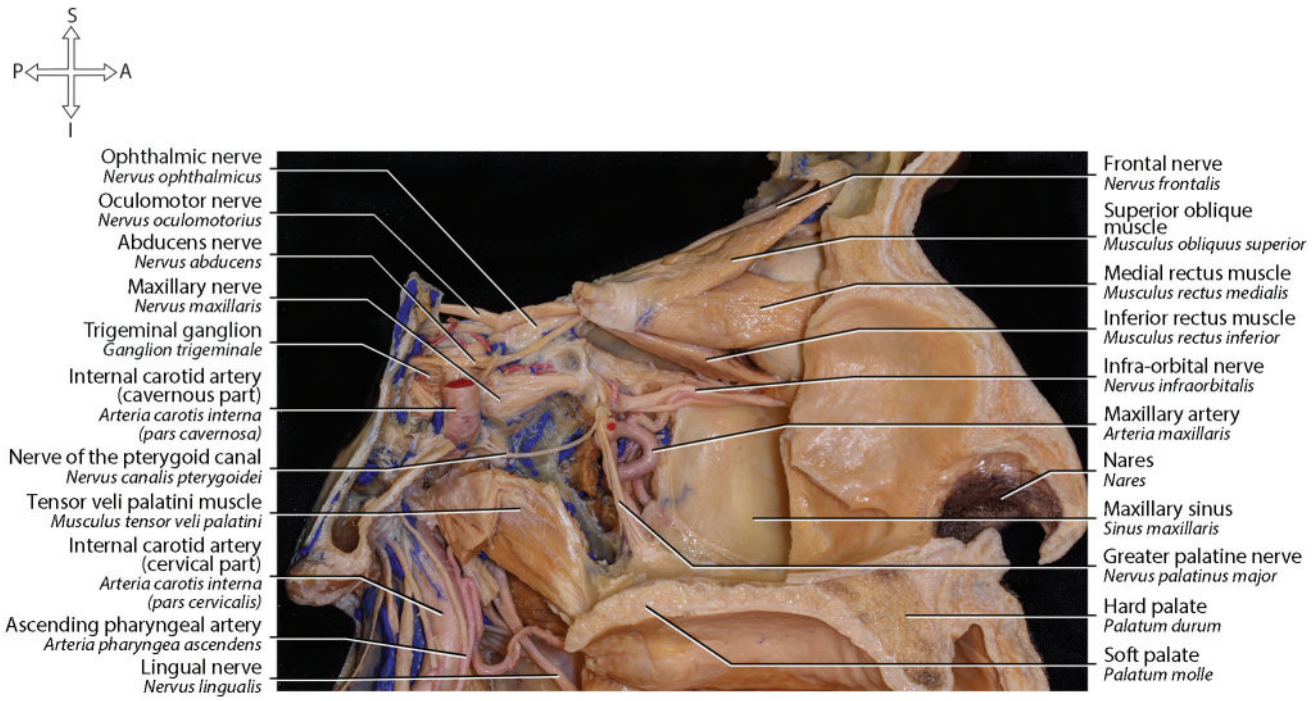

Fig. 3.5 Cadaveric dissection in the midsagittal plane, medial view of the left pterygopalatine fossa (PPF). A segment of the cavernous part of the internal carotid artery has been resected to expose the nerves in the cavernous sinus. The PPF has been opened and most of the pterygoid process removed. The auditory tube has been removed. (Reproduced with permission from Peris-Celda M, Martinez-Soriano F, Rhoton AL Jr., eds. Rhoton's Atlas of Head Neck and Brain. 2D and 3D Images. 1st ed. Thieme; 2017.)

Table 3.1 Classifications of juvenile nasopharyngeal angiofibroma

Sessions et al 1981[a]		Radkowski et al 1996[b]	
IA	Limited to posterior nares and/or nasopharyngeal vault	IA	Limited to nose and/or nasopharyngeal vault
IB	Extension into one or more sinuses	IB	Extension into one or more sinuses
IIA	Minimal extension through the sphenopalatine foramen into the pterygomaxillary fossa	IIA	Minimal extension into pterygomaxillary fossa
IIB	Full occupation of the pterygomaxillary fossa, displacing the posterior wall of the maxillary antrum, lateral/anterior displacement of maxillary artery branches, with or without erosion of orbital bones	IIB	Full occupation of the pterygomaxillary fossa with or without erosion of orbital bones
IIC	Infratemporal fossa into cheek	IIC	Infratemporal fossa with or without cheek or posterior to the pterygoid plates
III	Intracranial extension	IIIA	Erosion of the skull base—minimal intracranial extension
		IIIB	Erosion of the skull base—extensive intracranial extension with or without cavernous sinus invasion

[a]Ye L, Zhou X, Li J, Jin J. Coblation-assisted endonasal endoscopic resection of juvenile nasopharyngeal angiofibroma. J Laryngol Otol 2011; 125(9):940–944.
[b]Wormald PJ, Van Hasselt A. Endoscopic removal of juvenile angiofibromas. Otolaryngol Head Neck Surg 2003;129(6):684–691.

initial incision is made in the gingivolabial and gingivobuccal sulci and runs laterally to the maxillary tuberosities (**Fig. 3.6**). A transfixion incision is made along the nasal septum and is then extended around the limen vestibulae. The soft tissue of the nose is degloved up to the level of the infraorbital foramen, exposing the bony structures of the nose and maxilla below. The midfacial degloving approach provides access to the nasal cavity, nasopharynx, PPF, pterygomaxillary space, ITF, maxillary and sphenoid sinuses, cheek, and orbit. Risks of the midfacial degloving technique include facial paresthesia, nasal crusting, oroantral fistula, and epiphora.

If the tumor has significant involvement of the ITF and medial extension into the cavernous sinus, a LeFort I osteotomy approach can provide better exposure (**Fig. 3.7**). A fracture is made from the pyriform aperture to the pterygoid plates to detach the maxilla from the cranial base, thus allowing for exposure of the central skull base. This approach also involves resection of the posterior maxillary sinus wall, allowing for good visualization of the

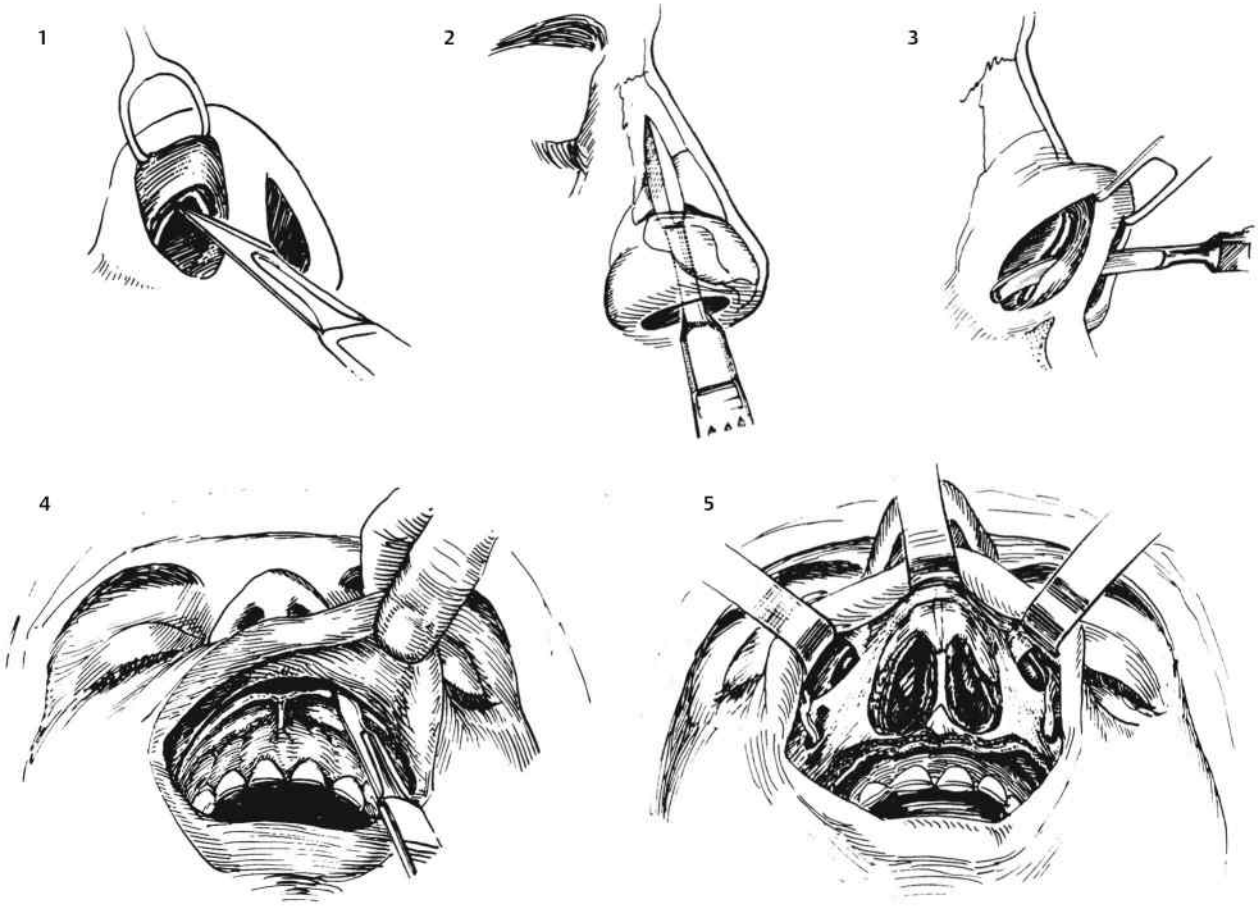

Fig. 3.6 Midface degloving. Midface degloving is accomplished by a combination of bilateral intranasal (1), intercartilaginous (2), septal transfixion (3), and sublabial (4) incisions followed by subperiosteal elevation of all soft tissue of the midface (5). (Reproduced with permission from Casson PR, Bonanno PC, Converse JM. The midfacial degloving procedure. Plast Reconstr Surg 1974;53:102–103.)

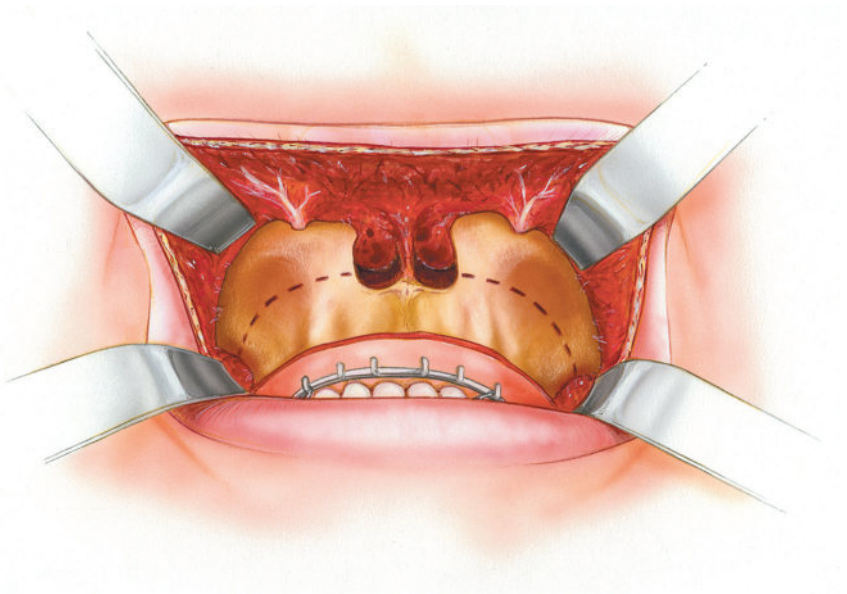

Fig. 3.7 LeFort I osteotomy. The nasal mucosa is carefully elevated. The *dashed line* represents the location of the Le Fort I osteotomy. (Reproduced from Fessler R, Sekhar L, eds. Atlas of Neurosurgical Techniques. Spine and Peripheral Nerves. 2nd ed. Thieme; 2016.)

vascular supply to maximize hemostasis. Risks of this approach include stunted vertical maxillary growth in developing children, dental denervation, aseptic necrosis of the maxilla, cerebrospinal fluid (CSF) leak, and extraocular nerve palsy.

For tumors involving the ITF, middle cranial fossa, and lateral cavernous sinuses, an ITF approach may be used. Given the wide exposure, the internal maxillary artery is identified and ligated early on for improved intraoperative hemostasis. Cosmetically, this approach minimizes facial scarring, and does not risk midface deformity. However, this approach is unable to reach tumors medial to the abducens nerve in the cavernous sinus, so additional medial approaches must be supplemented for those tumors.

Recent advances in skill and technology have made it possible to access the PPF and beyond with endoscopic technique and made endoscopy a viable option for JNA surgery (**Fig. 3.8**). Advantages to endoscopic JNA resection include decreased intraoperative time and shorter postoperative hospitalization stays. With endoscopic surgery, there is of course no skin incision, but it also minimizes bone removal, thus avoiding complications associated with open approaches including epiphora, trismus, and possible facial growth deformity. JNAs characteristically have a well-defined capsule which creates an optimal surgical plane for endoscopic dissection.

Major challenges of endoscopic sinus surgery include hemorrhage control, small operative field, and limited visualization. Intraoperative hemorrhage control can be addressed with the use of preoperative embolization, which can reduce intraoperative blood loss as much as 70%. The most commonly involved vessels include branches of the internal maxillary artery (e.g., sphenopalatine artery and vidian artery), ascending pharyngeal artery, mandibular artery, and facial artery, which can individually be targeted with superselective catherization techniques. Further improvement to control intraoperative blood loss came in the form of coblation technology. The coblator acts at a low temperature (60–70°C), allowing for it to ablate tumor tissue with less damage to adjacent tissues, while concurrently sealing feeding blood vessels. Recent studies have demonstrated the superiority of coblator-assisted endoscopic JNA resection in

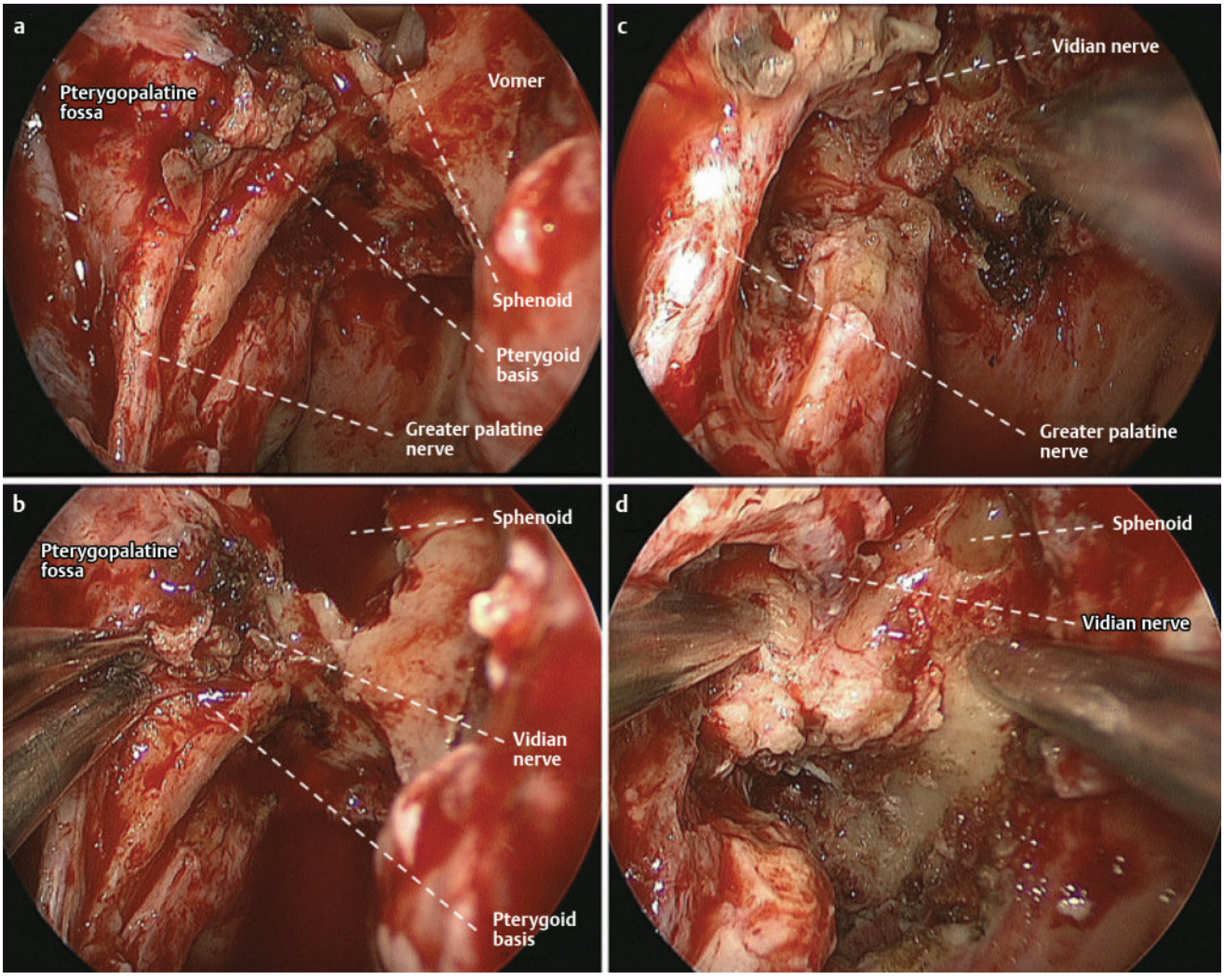

Fig. 3.8 The endoscopic transpterygoid approach (right side with a 0-degree endoscope). **(a)** After exposure of pterygopalatine fossa and greater palatine nerve. **(b)** Note the vidian canal opening with lateral mobilization of pterygopalatine fossa. **(c, d)** Vidian and greater palatine nerve preserved after the pterygoid base and the medial pterygoid plate were partially removed. (Reproduced from Pinheiro-Neto C, Fernandez-Miranda J, Prevedello D, et al. Transposition of the pterygopalatine fossa during endoscopic endonasal transpterygoid approaches. J Neurol Surg B Skull Base 2013; 74(5):266–270.)

decreasing blood loss as well as operative time, although studies are currently limited to tumors of Radkowski stage IIC or less.

Hemostasis and surgical access are further enhanced by switching from one-hand endoscopy to a two-surgeon technique. As first described, this technique involved removal of the bony septum and incision of the bilateral mucosal surfaces to allow for additional instruments to be placed via the nonoperative nostril into the operative field. The second surgeon retracted the tumor into the opposite nasal cavity to create space for the first surgeon to resect the tumor, and/or to provide additional suctioning in the event of intraoperative hemorrhage. A variation of this technique was later developed that used a more posterior transseptal perforation, which provides a wider operative field and decreases the need for piecemeal resection.

With the aforementioned enhancements, endoscopic techniques started to gain acceptance for use on JNAs. Although exposure is still restricted compared to open approaches, the use of an endoscope brings better lighting to the field and provides a magnified, multiangled view of the mass and its surrounding anatomy. Additionally, with integrated image-guidance technologies, the location of vital structures, such as the orbital apex, optic nerve, cavernous sinus, and carotid artery can be tracked. Recent studies showed equal efficacy between endoscopic and open approaches, even for high-stage tumors (up to IIIA) and with that, endoscopic techniques may well be applicable now for *all* JNAs except those with extensive intracranial extension.

Approach of Choice

The tumor in our patient was classified as Radkowski stage IIC, involving the nasal cavity, nasopharynx, maxillary, ethmoid and sphenoid sinuses, as well as the PPF, ITF, and orbital apex. Buoyed by the studies mentioned above which showed equivalency in open and endoscopic techniques for higher-stage tumors, we chose the endoscopic approach with the goal of total resection of his large tumor.

Questions

1. What risks would you mention to the patient during the informed-consent process, and what deficits would you anticipate after surgery?
2. Provided that the operation goes smoothly, when would you request your first postoperative imaging study?

■ Description of the Technique

Prior to tumor resection, the patient underwent a cerebral angiogram with preoperative embolization. On cerebral angiogram **(Fig. 3.9)**, significant blood supply was seen originating from the left maxillary artery. The left maxillary artery was subsequently embolized using Embosphere particles ranging from 300 to 500 µm and 500 to 700 µm, with immediate postembolization decrease in vascularization. The patient

The Three Approach Elements

Corridor: endonasal

Craniotomy: N/A

Modifier: access to ITF and PPF

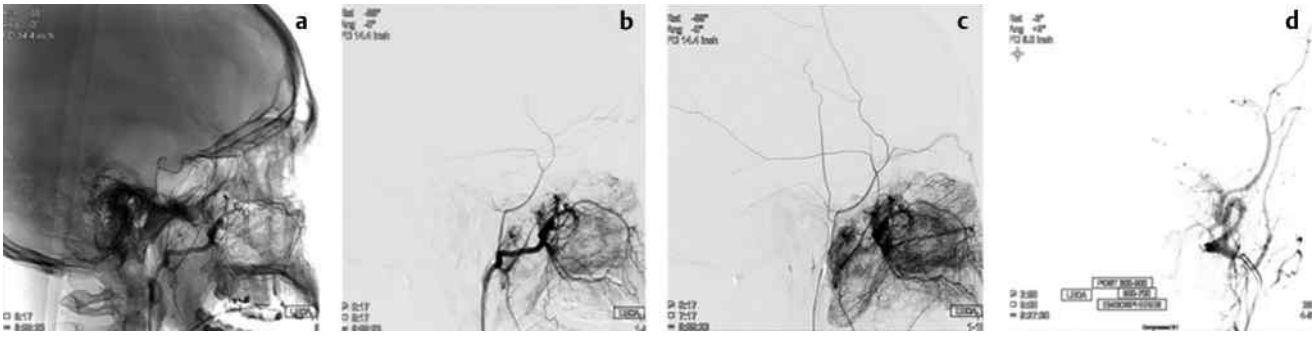

Fig. 3.9 Cerebral angiogram and embolization of sphenopalatine artery. (a) Sagittal view of the patient's pre-embolization cerebral angiogram showed a rich vascular network feeding the JNA, most prominently from the left maxillary artery. **(b-d)** The left maxillary artery was successfully embolized with embosphere particles with immediate decrease in vascularization.

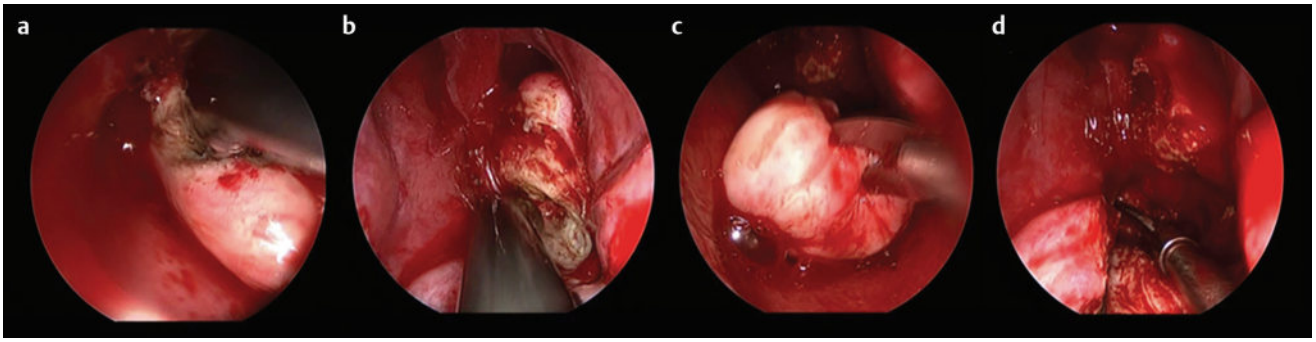

Fig. 3.10 Anterior tumor resection. The anterior portion of the tumor was removed using a combination of coblation **(a, b)** and blunt dissection **(c, d)** from the nasal cavity.

underwent endoscopic resection of JNA involving the nasal cavities, paranasal sinuses, PPF, ITF, orbital apex, and pterygoid plates (see **The Three Approach Elements).**

The patient was placed supine and draped in the standard fashion. Nasal endoscopy on the left revealed a large nasal mass obstructing the nasal vestibule. The Coblator surgical system was used to begin piecemeal resection of this massive JNA. Ablation of approximately the anterior one-third of tumor was performed with a combination of coblation, monopolar cauterization, and blunt and sharp dissection **(Fig. 3.10)**. Similar techniques were utilized to bisect the tumor horizontally and truncate the tumor in the nasal cavity from the maxillary,

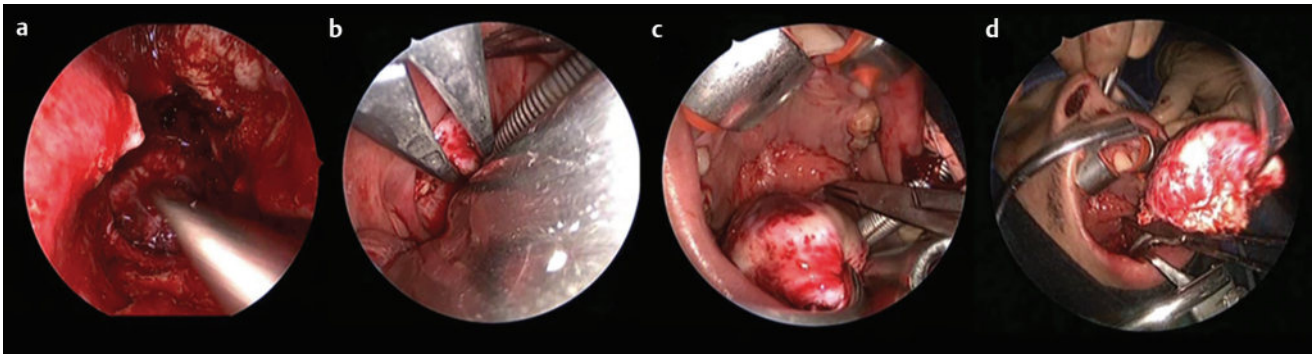

Fig. 3.11 Oral delivery of tumor. **(a)** The inferior and posterior portion of the tumor can be seen deep within the nasopharynx. **(b)** An oral retractor was placed and **(c, d)** the tumor was delivered via the oropharynx out of the oral cavity.

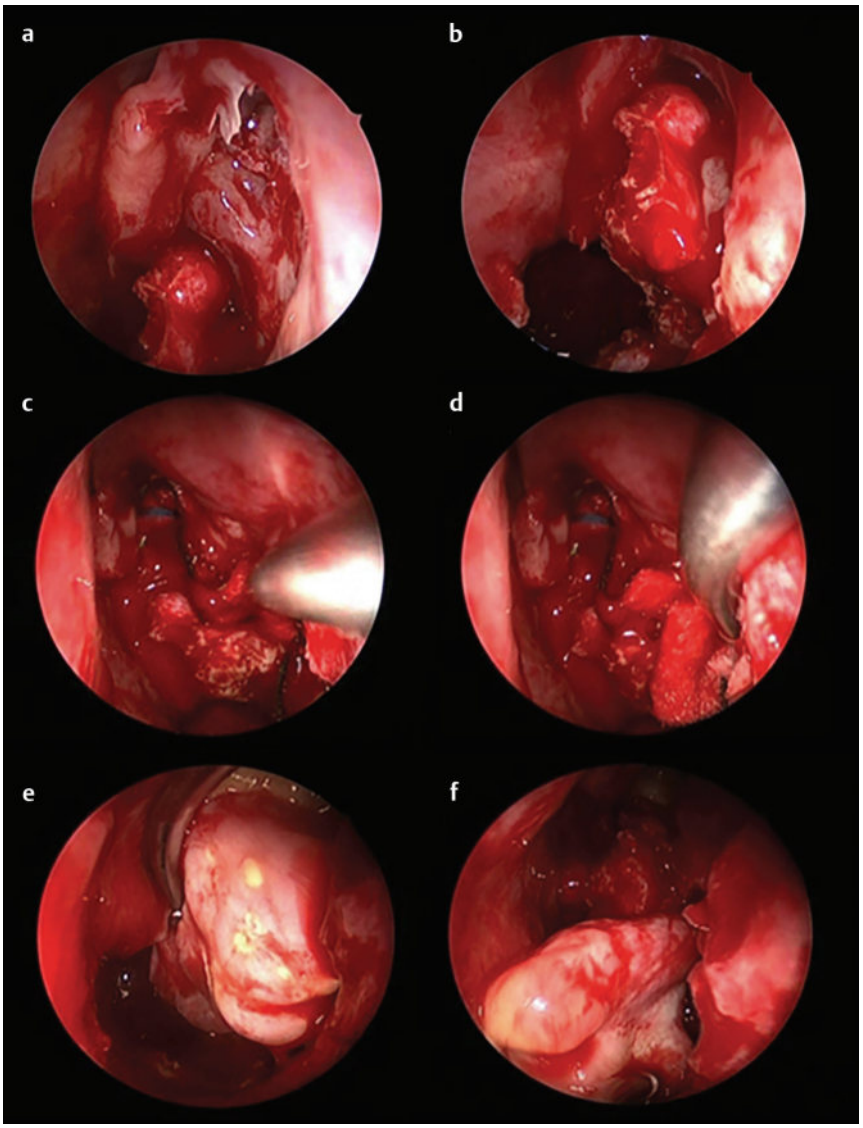

Fig. 3.12 Maxillary sinus delivery of tumor. **(a)** Tumor extension into the maxillary sinus. **(b)** Maxillary antrostomy was performed for further exposure, and **(c, d)** cottonoids were placed in the sinus to help deliver this component of the tumor via the nasal cavity **(e, f)**.

PPF, and ITF components. This was necessary to create space for lateral and superior dissection of the tumor. Bleeders from the ethmoidal vessels were addressed using monopolar cautery. Using similar instrumentation and techniques, the inferior portion of the tumor was resected to the nasopharynx, and sphenoid face. The posterior and inferior portion of the tumor was removed transorally given its size **(Fig. 3.11).** The tumor was encapsulated over the posterior portion of the mass.

Next, an endoscopic maxillectomy, along with a maxillary antrostomy, superior ethmoidectomy, and frontal sinusotomy was performed. The JNA was dissected out of the maxillary sinus using blunt dissection and placing cottonoids in the sinus to deliver this tumor component into the nasal cavity **(Fig. 3.12).** The medial PPF component in continuity to the superior ethmoid and maxillary components of the tumor was dissected using coblation and delivered through the nasal cavity **(Fig. 3.13)**. Using Kerrison rongeurs, the anterior wall of the PPF (equivalent to the medial portion of PWMS) was removed next, and the PPF and ITF component of the tumor abutting the orbital apex was dissected with sharp and blunt technique from the anterior fat and posterior neural compartments. Suction monopolar on a low setting, as well as blunt dissection under angled endoscopic visualization, were used to accomplish this task **(Fig. 3.14)**.

Using two-hand technique through the ipsilateral nostril, bipolar cautery, as well as a suction cautery, the ITF mass was resected. Multiple clips were placed on feeders to the mass

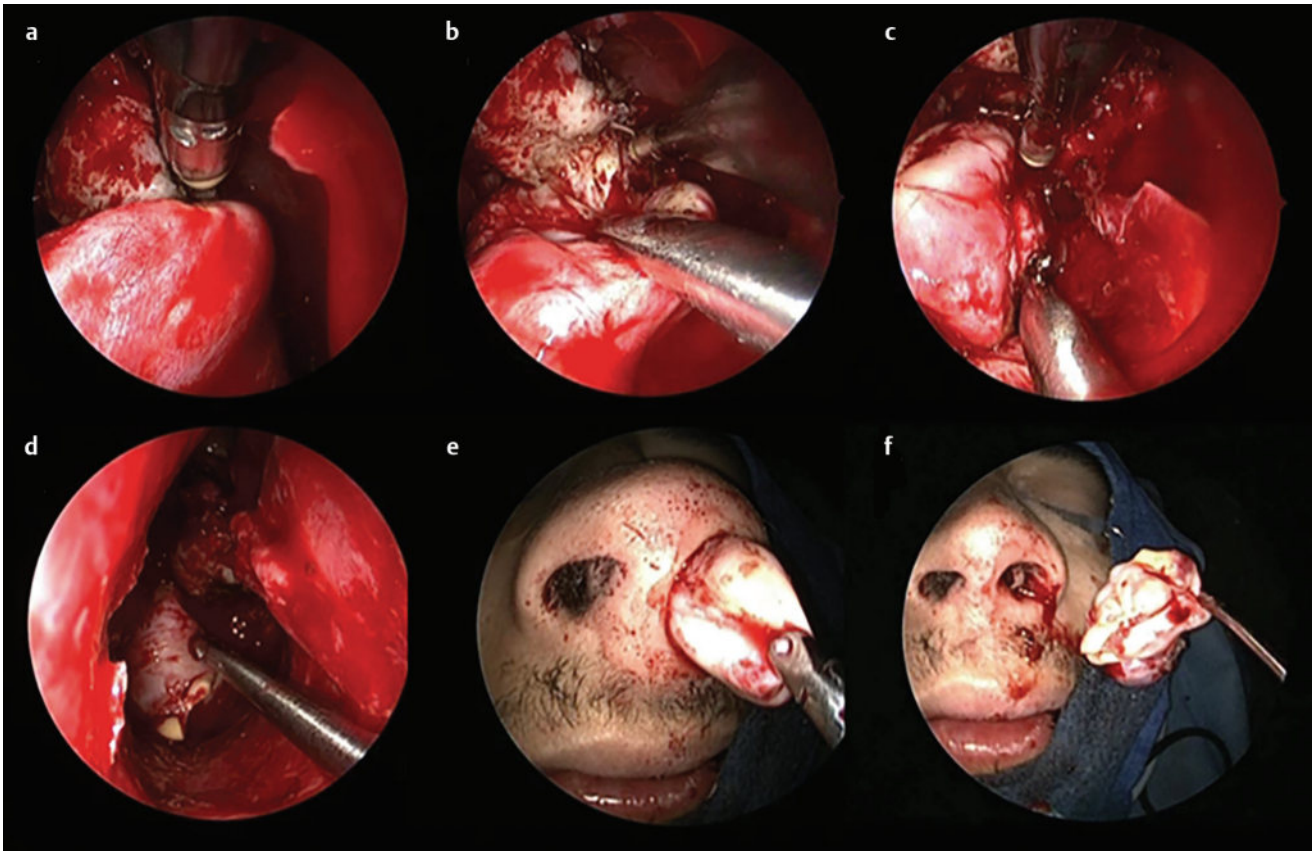

Fig. 3.13 Medial pterygopalatine fossa (PPF) portion of tumor dissection. Coblation was used to dissect the medial PPF component in continuity to the superior ethmoid and maxillary components of the tumor **(a–c)**, which was then delivered via the nasal cavity **(d-f)**.

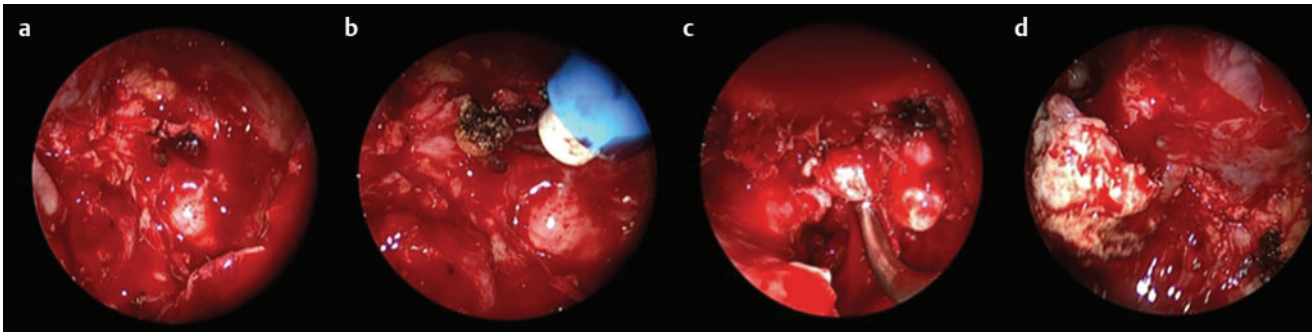

Fig. 3.14 Pterygopalatine fossa (PPF) and infratemporal fossa (ITF) portion of tumor dissection. Anterior wall of PPF was removed (not shown) to expose the PPF and ITF tumor components. **(a)** Suction monopolar and blunt dissection were used to carry out the dissection of tumor abutting the orbital apex **(b–d)**.

from the internal maxillary artery, which included the posterior superior alveolar artery, descending palatine arteries as well as sphenopalatine arteries. Once the ITF of the mass was resected, a margin was obtained and sent for pathologic examination. Large pieces of Gelfoam and Surgiflo were placed on the PWMS, PPF, and sphenoid regions.

A postoperative MRI was obtained, and a tumor residual involving the orbital apex, lateral recess of the sphenoid sinus, and ITF was discovered (**Fig. 3.15a–c**). We concluded that inadequate exposure of the sphenoid sinus and lateral sphenoid wall was the cause of the problem, and as such, the patient was scheduled for a repeat operative procedure 5 days after the first surgery. A wide sphenoidotomy with extensive exenteration of the sphenoid sinus was performed. Kerrison rongeurs were used to thoroughly expose the lateral recess of the sphenoid sinus. Two small dumbbell-shaped tumor components were resected from the lateral recess of the sphenoid sinus interdigitating with the pterygoid plates, medial contents of the PPF, and the ITF. The vidian nerve and V2 were sacrificed, while V3 was spared, to achieve a complete resection. Cautery and clips were applied for hemostasis. Hemostatic agents were placed in the surgical cavity as this second-stage operation concluded.

Surgical Pearls

1. Although a biopsy was done for this patient, in the vast majority of JNAs, this potentially bloody procedure can be avoided.
2. The use of two-surgeon, four-hand technique is critical for success in the resection of such a complex and extensive JNA. It improves hemostasis, maximizes surgical access, and minimizes operative time. While as a regular operative partner in endoscopic procedures the neurosurgeon may seem to have no role in this tumor resection, she or he may be an appropriate assistant in the two-surgeon paradigm.
3. Although this operation turned into a staged operation, routine staging of these procedures can be avoided by aggressively widening the surgical access from the beginning. A sphenoidotomy at the first operation would have likely eliminated the need for this patient's second operation.

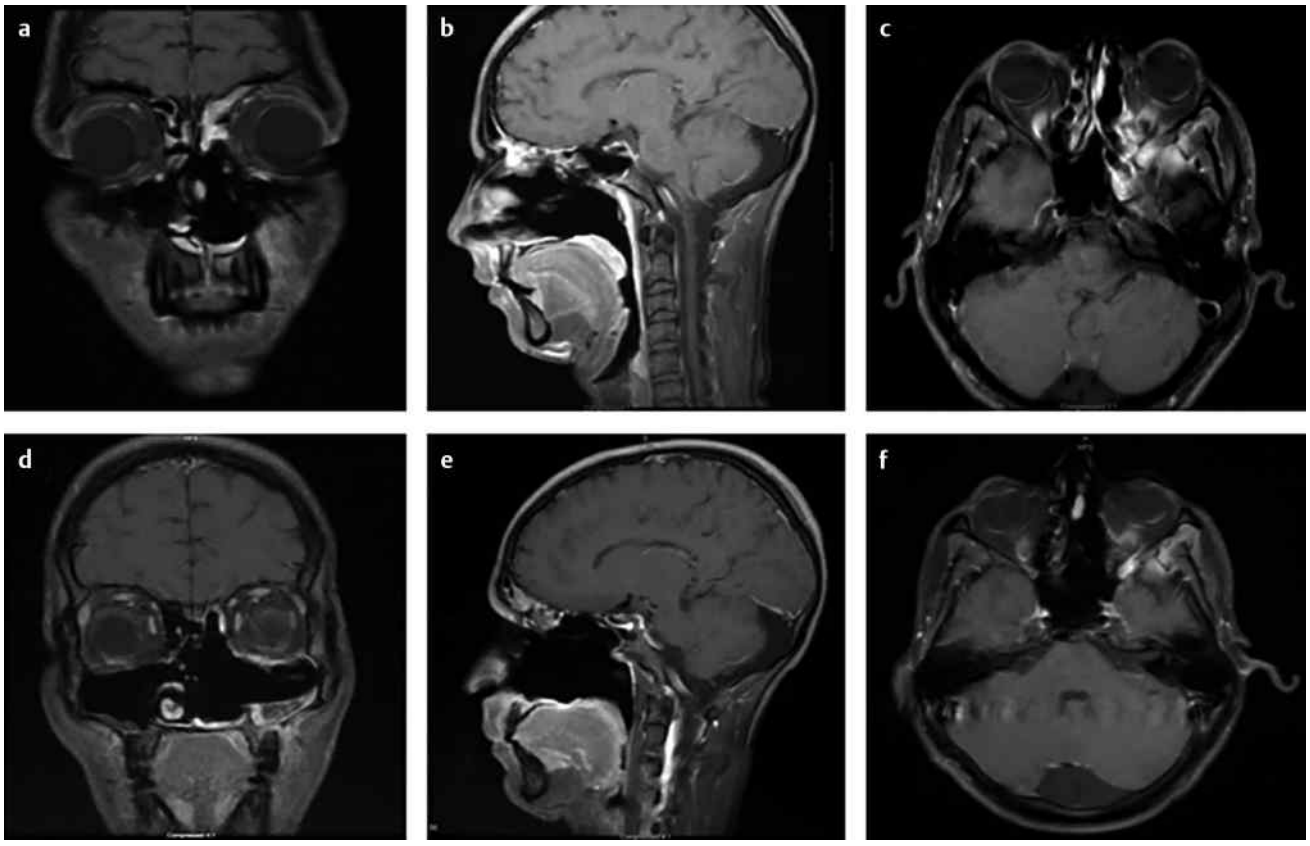

Fig. 3.15 Postoperative MRI scan. (a–c) Immediate postoperative MRI scans and **(d–f)** revision postoperative MRI scans**. (a)** Coronal, **(b)** sagittal, and **(c)** axial T1-weighted postcontrast MRI images demonstrated residual tumor in the lateral sphenoid sinus, infratemporal fossa, and orbital apex. **(d–f)** Comparable MRI images obtained after revision surgery demonstrated complete tumor removal and normal postoperative changes.

Aftercare

The patient returned to clinic for his first follow-up visit within 2 weeks of surgery. Left-sided numbness along the cranial nerve V2 distribution was noted on examination, as expected after intraoperative nerve sacrifice. No trigeminal nerve (V3) deficit or visual changes were observed. The patient also denied postoperative epistaxis, and his residual nasal congestion and crusting were eventually resolved with consistent nasal saline spray use. An MRI after surgery demonstrated expected postoperative changes without residual tumor (**Fig. 3.15d–f**). The patient had consistent follow-up in clinic, and a 2-year follow-up MRI showed no evidence of residual or recurrent disease and resolving postoperative changes.

Close postoperative follow-up with imaging can help detect residual disease and/or recurrence. Early postoperative CT imaging of patients with Radkowski stage III tumors obtained within 5 days of surgery has a specificity of 83%, indicating a high reliability in detecting true residual disease. However, imaging obtained any earlier than 72 hours may have limited value due to confounding by postoperative inflammatory changes. Patients who are found to have residual disease on imaging must undergo careful evaluation regarding further management. Small tumor remnants in asymptomatic patients may be observed with serial imaging, as they have been shown to stabilize or regress spontaneously.

Although the exact timing of additional postoperative imaging is not clear, one study done on endoscopically resected JNAs recommends CT scans 6 to 12 weeks after resection, followed by a second CT scan performed 4 to 6 months postoperatively to identify recurrent tumor. Overall JNA recurrence rates range from 22 to 37.5%, although the exact rate is highly dependent on preoperative tumor staging, rate of growth, and degree of surgical resection. Higher recurrence rates are seen in JNAs that involve the ITF, sphenoid sinus, pterygoid plates and clivus, cavernous sinus, foramen lacerum, and anterior fossa. Proper identification, meticulous subperiosteal dissection, and drilling of these areas may successfully prevent future recurrence.

Perspective

Hussam Abou-Al-Shaar, Wayne D. Hsueh, Jean Anderson Eloy, and James K. Liu

■ Introduction

The authors of the previous section have eloquently described their skillful use of endonasal endoscopic techniques to remove a large JNA, avoiding transfacial surgery and its multiple risks. However, despite the advancement of technique, improvement of equipment, and accumulation of experience, endoscopic endonasal approaches still have inherent limitations. One such limitation is insufficient access and exposure of large JNA with multicompartmental extension to lateral locations in the skull base. For such large tumors, which extends well into the ITF, possibly reaching its lateral extreme, additional exposure can be obtained if multiple endoscopic corridors are utilized in combination with each other. We discuss the application of such a strategy in this section.

■ Case Presentation

An otherwise healthy 13-year-old boy presented to our service complaining of nasal obstruction for the past 6 months. His neurological examination was unremarkable. Endoscopic examination of both nasal cavities demonstrated a nasopharyngeal mass extending into the right nasal cavity. CT scan and MRI depicted a large mass in the nasopharynx extending bilaterally into both nasal cavities, bilateral sphenoid sinuses, right PPF, ITF, and right inferior orbital fissure. Erosion was also evident in the anterior clivus and pterygoid plate (**Fig. 3.16**).

■ Diagnosis and Assessment

The constellation of symptoms including nasal obstruction, epistaxis, and a nasopharyngeal mass in an adolescent male should raise the suspicion for the diagnosis of JNA. These patients should undergo further evaluation and definitive surgical management by a multidisciplinary skull base team. All patients with JNA should receive a high-resolution CT and MRI scans in order to evaluate the bony architecture, delineate the tumor location and extension, and plan the surgical approach.

For reasons already discussed in the previous section, biopsy is usually unnecessary in the setting of a typical presentation, but if an atypical history or presentation warrants a tissue diagnosis, the procedure should be performed in an operating room under general anesthesia due to the high vascularity of the tumor and the potential of life-threatening bleeding. The various staging systems for JNA have also been discussed, and the most commonly utilized systems include the Radkowski, Andrews–Fisch, and Snyderman (UPMC) staging systems, among others. These staging systems classify JNAs primarily based on their location and tumor extension to aid surgeons in planning the surgical approach.

Since JNA is a hypervascular tumor, embolization of the blood supply prior to surgical resection is commonly recommended (**Fig. 3.17**). In our practice routine, the angiogram and endovascular procedure is done within 24 to 48 hours before surgery to prevent revascularization of the tumor, which can take place rapidly. We utilize polyvinyl alcohol particles (diameter between 150 and 200 µm) to embolize the feeders, and have found that this makes intraoperative blood loss much more manageable during tumor resection. The primary arterial blood supply of JNA comes from the internal maxillary artery. It is important to rule out the presence of shunts with the ICA or vertebrobasilar system during diagnostic angiography, as embolization in these scenarios could lead to a catastrophic stroke. It is also important to recognize arterial feeding vessels that arise from the ICA circulation, which are usually seen in larger and giant JNAs with intracranial extensions. These ICA feeders typically cannot be safely embolized, making resection much more challenging for these tumors.

Fig. 3.16 Preoperative images. T1-weighted post-gadolinium MR image; **(a)** sagittal, **(b)** coronal, and **(c)** axial views showing a JNA involving bilateral nasal cavity, right pterygopalatine fossa, and far-lateral infratemporal fossa.

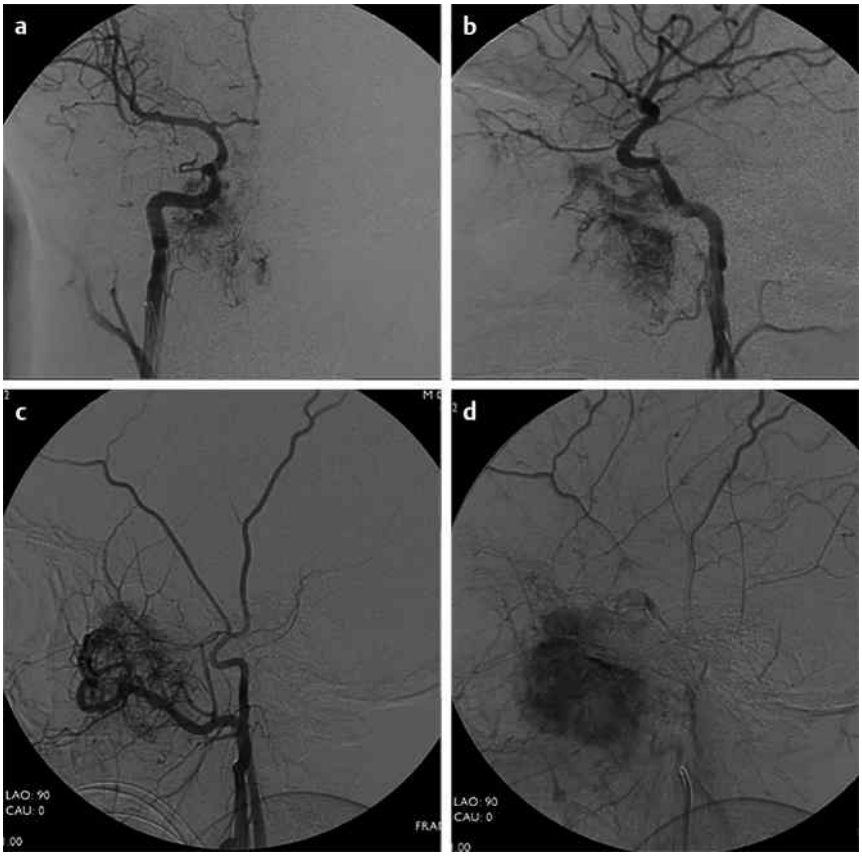

Fig. 3.17 Preoperative angiogram. Right internal carotid injection, **(a)** anteroposterior view, and **(b)** lateral view showed hypervascularity of the JNA with arterial supply fed from the right cavernous internal carotid artery. **(c, d)** Right external carotid artery injection and lateral views showed supply from the right internal maxillary arteries. Only the arterial feeders from the external carotid artery could be safely mobilized prior to surgery.

■ Anatomical and Therapeutic Considerations

The patient in our case had a typical presentation of JNA. Given his age, as well as his clinical and radiological presentation, we elected to proceed with surgical resection of his lesion with an aim of achieving a gross total resection. As previously mentioned, multiple classification and staging schemes have been developed to aid the surgeon in planning and choosing the optimal surgical approach. Whichever scheme one chooses, it is of paramount importance to study the radiological studies in detail to plan the operative approach accordingly.

The large tumor in our example was centered in the nasopharynx and extended bilaterally into nasal cavities, bilateral sphenoid sinuses, the right PPF, ITF, and right inferior orbital fissure. Given the lateral extension of the tumor, a purely endonasal endoscopic approach (EEA) may not provide adequate exposure to remove the tumor in an efficient manner. Alternatively, open transfacial approaches can potentially be utilized to access the tumor, and these include the lateral rhinotomy, midfacial degloving, Le Fort I osteotomy, transcervical, and facial translocation approaches. However, these approaches are associated with limited visualization, lack of adequate illumination, and many facial cosmetic complications such as facial growth retardation, malocclusion, transfacial scars, and facial nerve injury.

Approach of Choice

Given the multicompartment location of the tumor, we elected a combined endoscopic graduated multiangle, multicorridor approach to remove his JNA. This combined endoscopic-assisted technique allows one to fully access the tumor in all compartments and obviates the need for open transfacial approaches with their associated complications. We have utilized these approaches in a variety of tumors extending into various compartments with excellent outcomes. **Table 3.2** outlines the surgical corridors, approaches, and the locations that can be accessed using these endoscopic graduated multiangle, multicorridor approaches. Since the specific tumor in our case example occupied the nasopharynx on both sides, with significant lateral extension into the right ITF, we elected to use a combined EEA–sublabial (Caldwell–Luc) maxillotomy approach, using both the binostril–endonasal, as well as the sublabial–transmaxillary corridors (**Fig. 3.18**).

■ Description of the Technique

After preoperative embolization, the patient was taken to the operating room where general anesthesia was established with controlled hypotension (mean arterial pressure goals of 60–70 mm Hg). The patient was placed supine with 15 to 20 degrees of reverse Trendelenburg to decrease the central venous pressure. Neurosurgical pledgets (3 × 0.5 in) soaked with 10 mL of 1:1,000 epinephrine were placed inside the nose for decongestion. The root of the middle turbinate, lateral nasal wall, and nasal septum were injected with 1% lidocaine with 1:100,000 epinephrine.

The tumor in the nasopharynx was exposed on both sides, using a binostril endoscopic endonasal approach with a septostomy to allow bimanual binostril access (**Fig. 3.19**). Tumor

Table 3.2 Site-specific targets of graduated multicorridor approach

Surgical corridor(s)	Approaches	Locations that can be accessed
Uninostril	Unilateral medial maxillectomy, sphenoidotomy, ethmoidectomy, transpterygoid	Ipsilateral nasal cavity, sphenoid sinus, ethmoid sinus, frontal sinus, hemi-skull base (cribriform), maxillary sinus, pterygopalatine fossa, posterior nasopharynx
Binostril	Bilateral sphenoidotomy, septectomy, bilateral ethmoidectomy, bilateral frontal sinusotomy, unilateral medial maxillectomy, transpterygoid	Bilateral nasal cavity, sphenoid sinuses, ethmoid sinuses, frontal sinuses, entire cribriform plate, ipsilateral maxillary sinus, pterygopalatine fossa, posterior nasopharynx, medial infratemporal fossa
Binostril + Caldwell–Luc	Bilateral sphenoidotomy, septectomy, bilateral ethmoidectomy, bilateral frontal sinusotomy, unilateral medial maxillectomy, transpterygoid, sublabial maxillotomy	Bilateral nasal cavity, sphenoid sinuses, ethmoid sinuses, frontal sinuses, entire cribriform plate, ipsilateral maxillary sinus, pterygopalatine fossa, posterior nasopharynx, medial and lateral infratemporal fossa
Binostril + Caldwell–Luc + transcranial	Bilateral sphenoidotomy, septectomy, bilateral ethmoidectomy, bilateral frontal sinusotomy, unilateral medial maxillectomy, transpterygoid, sublabial maxillotomy, orbitozygomatic craniotomy	Bilateral nasal cavity, sphenoid sinuses, ethmoid sinuses, frontal sinuses, entire cribriform plate, ipsilateral maxillary sinus, pterygopalatine fossa, posterior nasopharynx, medial and lateral infratemporal fossa, intracranial, cavernous sinus

Source: Reproduced with permission from Liu JK, Husain Q, Kanumuri V, Khan MN, Mendelson ZS, Eloy JA. Endoscopic graduated multiangle, multicorridor resection of juvenile nasopharyngeal angiofibroma: an individualized, tailored, multicorridor skull base approach. J Neurosurg 2016;124(5):1328–1338.

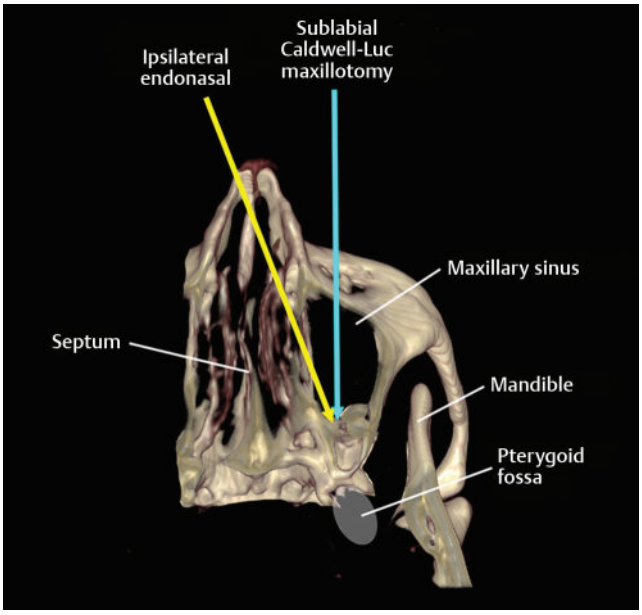

Fig. 3.18 Anatomy of the inferior lateral skull base (left side). The blue arrow shows the trajectory of the sublabial Caldwell–Luc maxillotomy approach. The yellow arrow shows the trajectory of the endonasal approach.

in the ITF was exposed using an ipsilateral endoscopic medial maxillotomy (maxillary antrostomy, inferior turbinectomy) from the right side, and also a right-sided sublabial Caldwell–Luc maxillotomy via a sublabial incision (**Fig. 3.19b**). During the Caldwell–Luc maxillotomy, care was taken to avoid injury to the infraorbital nerve superiorly and teeth roots inferiorly. In total, three portals of access were created (ipsilateral nostril, contralateral nostril, ipsilateral sublabial maxillotomy) to allow multiportal, multicorridor access. The endoscope was used in all three corridors to obtain the best visualization for tumor removal.

For these far laterally positioned JNAs, addition of the sublabial maxillotomy corridor allows more direct anterior access to the far-lateral portion of the ITF and excellent surgical freedom when mobilizing surgical instruments (**Fig. 3.20**). In this patient, maximal surgical freedom was achieved while placing the endoscope in the right nostril and introducing instruments through the left nostril and right maxillotomy corridors. We also prefer to use the 30-degree endoscope during skull base procedures because it provides increased angles of visualization in a multidirectional fashion. Rotating the 30-degree endoscope can provide optimal visualization to the maxillary sinus, ITF, anterior skull base, nasal floor, and posterior nasopharynx. Moreover, in the anterior maxillotomy corridor, the 30-degree endoscope can be angled medially to visualize the maxillary antrostomy and nasal septum.

The tumor was efficiently removed with a rotation–suction microdebrider. Special care was taken to avoid injury to the orbit and ICA with this device. Thus, it is important for the surgeon to be cognizant of the anatomical compartment they are working in and remain in "safe zones" (nasopharynx, nasal cavity, ITF) when using the microdebrider. Sequential bipolar cautery and early takedown of the internal maxillary artery was paramount for early devascularization of the tumor, even with preoperative embolization. Bimanual microsurgical technique must be used for safely dissecting the tumor away from important neurovascular structures (**Fig. 3.19d–f**). In this patient, there was no intraoperative CSF leak, as in most cases of extracranial JNAs. However, if an intraoperative CSF leak is detected, one should be prepared to reconstruct the skull base dural defect with a multilayered technique with a vascularized pedicled nasoseptal flap.

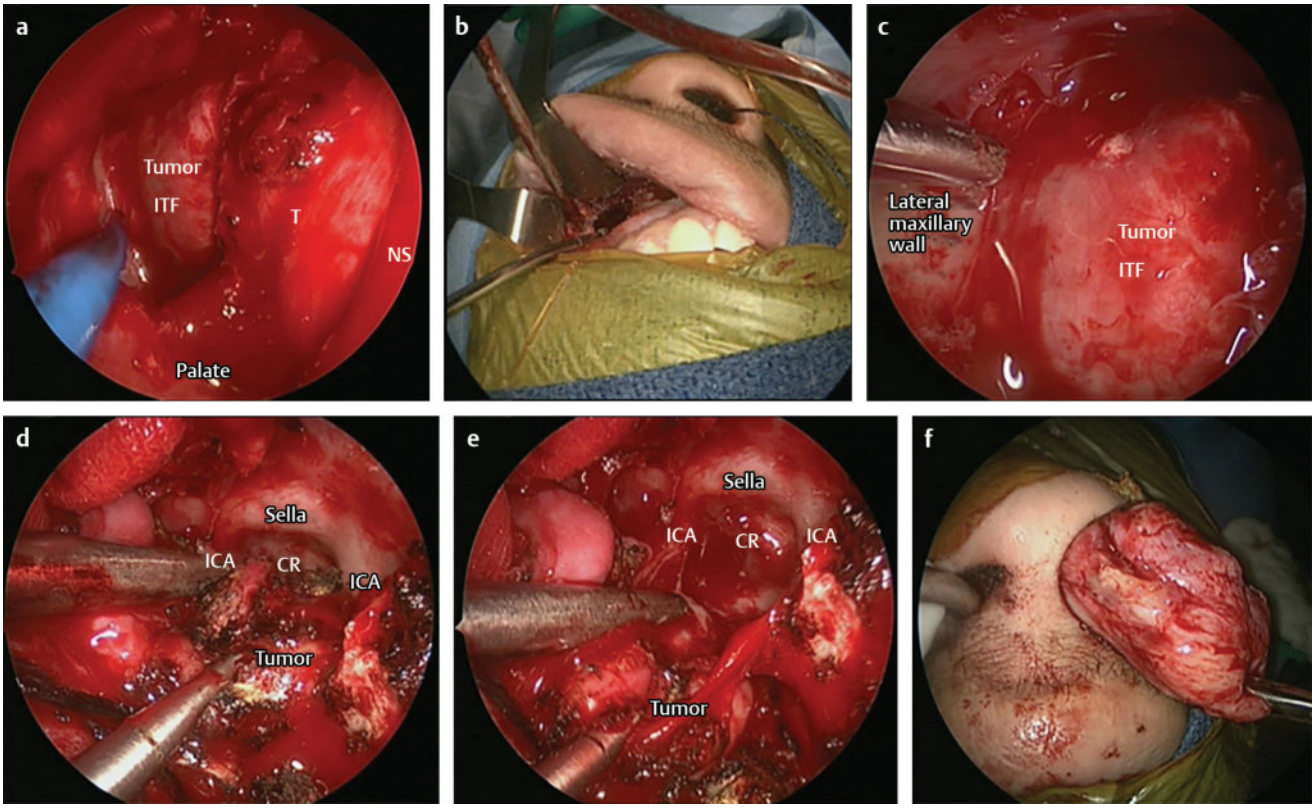

Fig. 3.19 Intraoperative endoscopic views. (a) A 30-degree endoscope was placed in the nasal cavity looking laterally to the right through the modified medial maxillotomy corridor. The tumor (T) in the nasal cavity and the tumor in the infratemporal fossa (ITF) was visualized. **(b)** A right Caldwell–Luc maxillotomy was performed with a sublabial incision. **(c)** The endoscope was placed into the sublabial maxillotomy to obtain a more direct view of the tumor in the ITF and the lateral maxillary wall. **(d–f)** The tumor in the sphenoid sinus was carefully peeled off of the skull base underneath the sella from the clival recess (CR). Care was taken not to injure both paraclival internal carotid arteries (ICA). The tumor was initially debulked with a microdebrider until it was small enough to deliver from the nostril **(f)**.

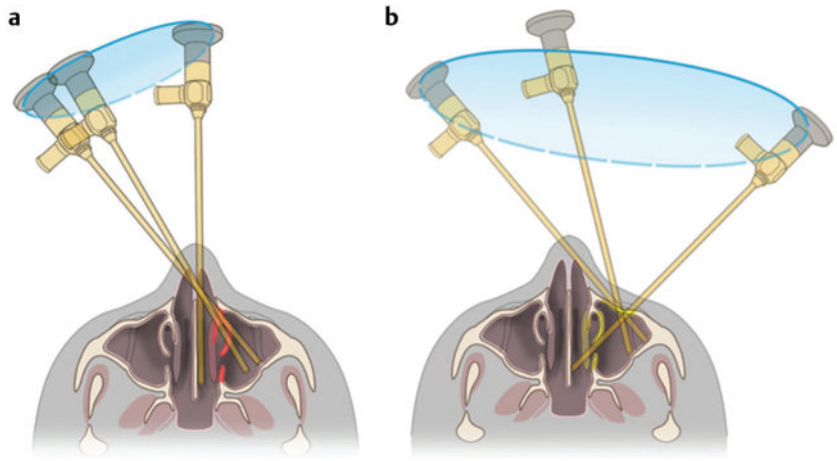

Fig. 3.20 Increased surgical freedom from sublabial Caldwell–Luc maxillotomy. The oval area is the surgical freedom for **(a)** the endonasal approach and **(b)** the sublabial approach. In this patient, maximal surgical freedom was achieved while placing the endoscope in the right nostril and introducing instruments through the left nostril and right maxillectomy corridor. (Reproduced from Elhadi A, Almefty K, Mendes G, et al. Comparison of surgical freedom and area of exposure in three endoscopic transmaxillary approaches to the anterolateral cranial base. J Neurol Surg B Skull Base 2014;75(5):346–353.)

■ Aftercare

The patient tolerated the procedure well. We did not use a lumbar drain postoperatively, per our routine. The patient was neurologically intact without complications and was discharged home on postoperative day 4. Because of the radiation exposure during embolization, the patient later developed transient geometrical alopecia that was managed conservatively. On his last follow-up appointment, 6 months after surgery, the patient was doing well without any evidence of residual tumor or recurrence (**Fig. 3.21**). Moreover, his geometrical alopecia had completely recovered.

Because of the high rate of JNA recurrence, it is recommended to follow these patients for a minimum of 5 years

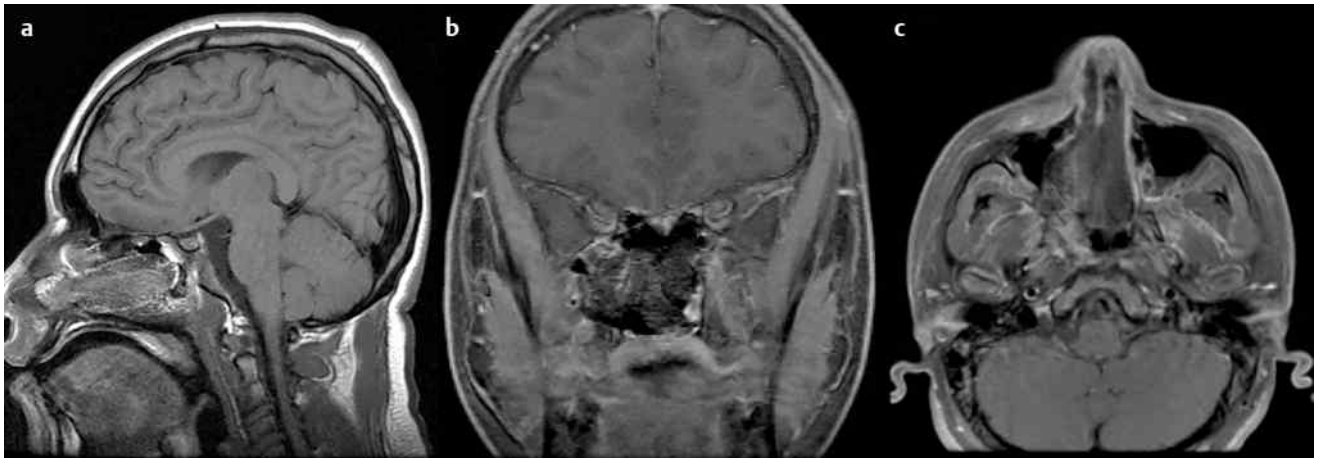

Fig. 3.21 Postoperative images. T1-weighted post-gadolinium MR images. **(a)** Sagittal, **(b)** coronal, and **(c)** axial views showed complete tumor removal without evidence of residual tumor.

after surgery. In our practice, we recommend a follow-up nasal endoscopy 3 months after surgery, and every 6 months after that for the first 3 years. Similarly, we recommend follow-up MRIs at 3 months after surgery, and once yearly thereafter for lifetime follow-up. If recurrence occurs, various treatment modalities can be instituted including reoperation, stereotactic radiosurgery, conventional radiotherapy, or watchful waiting. Some tumors may regress or stabilize when patients reach adulthood. However, this is not always the case, and therefore, continued radiologic follow-up is warranted.

Commentary

The case described in this section elucidates the importance of individualized approaches for managing JNAs. Traditional open transfacial approaches are rarely used in clinical practice nowadays because of their limited visualization, high morbidity profile, and poor aesthetic outcomes, especially in this young patient population. With the advancements of surgical techniques and instrumentations, endoscopic surgery for JNA has better outcomes, lower complications, less blood loss, and superior aesthetic outcomes compared to open transfacial approaches. However, despite its excellent clinical and safety profile, EEAs have their inherent limitations and complications. The case presented in this chapter demonstrated one such limitation. The EEAs remain limited in the management of laterally extending tumors occupying multiple compartments. The JNA in our patient extended laterally into the ITF and inferior orbital fissure, and utilizing a purely endonasal approach would prove challenging in dissecting the tumor from the far-lateral component of the ITF. In these cases, addition of the Caldwell–Luc maxillotomy allows multiportal, multicorridor access to this region with direct anterior access. This corridor provides better visualization, increased surgical freedom, and less trauma to the nasal structures.

In complex JNAs with intracranial extension, cavernous sinus involvement, and vascular supply from ICA feeders, we recommend adding a transcranial approach from above (orbitozygomatic transcavernous approach, in most cases) to the endonasal approach from below. The orbitozygomatic transcavernous approach from above allows the surgeon to safely peel the tumor away from the cavernous sinus and provide early vascular control of the cavernous ICA. Once this is established, the remainder of the tumor can be removed from the endonasal and transmaxillary corridors from below. In our experience, this combined approach has been very useful in managing these larger complex JNAs.

We favor an individualized, tailored, endoscopic graduated multiangle, multicorridor skull base approach for the resection of extensive JNA (**Table 3.2**). These endoscopic endonasal and endoscopic-assisted approaches provide wide access to tumors extending into different compartments that cannot be removed completely using a single endoscopic corridor and provide superior aesthetic outcomes compared to traditional transfacial approaches. The endoscope can also be utilized in traditional microsurgical corridors to visualize the tumor and aid in the resection process. These two-surgeon four-handed techniques are making major open transfacial surgery, with its risk of facial deformity and scarring, nearly obsolete.

In our case example, the JNA extended to the far-lateral portion of the ITF. Thus, a combined binostril EEA and ipsilateral endoscopic Caldwell–Luc maxillotomy provided three different corridors to fully access the tumor including the ITF component. Denker's anteromedial maxillotomy is an alternative approach to Caldwell–Luc maxillotomy that can be utilized in such patients. However, we prefer the Caldwell–Luc maxillotomy approach, as Denker's anteromedial maxillotomy has a higher risk of cosmetic nasal deformity and nasolacrimal duct obstruction.

Routine use of preoperative angioembolization in these patients is of a paramount importance, as it provides excellent delineation of the tumor vasculature and significantly reduces intraoperative bleeding (by 70%), making complete tumor removal easier and safer. Despite embolization, however, early intraoperative ligation and sectioning of the ipsilateral internal maxillary artery in the ITF is important to further devascularize the tumor. Once this is achieved, tumor removal is much easier to perform. Finally, all patients with JNA should undergo routine, long-term radiologic follow-up, even if the initial resection is complete. It is not uncommon for the tumor to recur in a delayed fashion.

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Part II

Tumors of the Anterolateral Skull Base

4 Anterior Clinoid *49*

5 Juxtasellar Cisterns *62*

6 Cavernous Sinus *73*

4 Anterior Clinoid

Michael C. Huang and Walter C. Jean

Keywords: anterior clinoid process, clinoidectomy, supraorbital keyhole, endoscope, minipterional craniotomy

■ Case Presentation

A 57-year-old man with chronic obstructive pulmonary disease (COPD) and asthma presented to our clinic with a 1-month history of severe right-sided headaches. He described the headaches as sharp, as if there was an "icepick" stuck in the right frontal region. He reported intermittent blurry vision in the right eye but denied any diplopia. On neurological examination, his vision in his right eye was 20/25 and 20/20 in the left eye. His cranial nerve, motor, and sensory examinations were unremarkable. Patient brought computed tomography (CT) and magnetic resonance imaging (MRI) images with him to his clinic visit. These showed a 4.3 × 3.3 × 4.7 cm mass lesion on the right side (**Fig. 4.1**).

Questions

1. What is your differential diagnosis based on the MRI findings?
2. What is the origin of the tumor? Where is it anchored anatomically?
3. What other studies should be considered to complete the work-up?

■ Diagnosis and Assessment

The MRI images showed a large extra-axial tumor with homogenous enhancement with gadolinium administration. The tumor was centered around the right anterior clinoid process (ACP) with extension along the planum sphenoidal and right sphenoid wing into the right anterior and middle cranial fossa, respectively (**Video 4.1**). The tumor extended to the right orbital apex and contacted the right optic chiasm and optic nerve without significant displacement. There was adjacent T2 hyperintensity in the adjacent parenchymal, suggestive of cerebral edema.

The ACP is the medial terminus of the lesser sphenoid wing. It is attached to the body of the sphenoid body medially by two roots: the anterior/superior root extends from the ACP and forms the roof of the optic canal, whereas the posterior/inferior root extends from the inferomedial aspect of the ACP and forms the optic strut (**Fig. 4.2**). Together with the optic strut and carotid sulcus, the ACP forms a bony collar around the carotid artery. Laterally, the ACP extends as the lesser wing of the sphenoid, forming the roof of the superior orbital fissure.

Because of its location at the junction of the anterior and middle cranial fossae, the anterior clinoid is frequently involved in meningiomas arising from both cranial fossae. Historically, meningiomas arising from the ACP have been variously described as medial sphenoid wing or suprasellar meningiomas. However, meningiomas of the anterior clinoid have distinctive behaviors that pose unique surgical challenges. Unlike medial sphenoid meningiomas, which grow anteriorly into the middle cranial fossa with frequent cavernous sinus involvement, anterior clinoidal meningiomas tend to grow cranially or into the optic foramen and only infrequently invade the cavernous sinus. The growth pattern of the tumor presented here is more consistent with a clinoidal meningioma than a sphenoid wing meningioma. There was also hyperostosis of the ACP, better appreciated on the CT scan (**Fig. 4.1a)**.

A preoperative cerebral angiogram was also performed. This provided detailed vascular anatomy in relationship to the tumor and demonstrated the degree of vascularity of the tumor. Often, large arterial feeders to the tumor may be embolized preoperatively to reduce intraoperative blood loss. In this case, there was minimal tumor blush, and therefore, no embolization was attempted.

■ Anatomical and Therapeutic Considerations

There are several critical neurovascular structures adjacent to the ACP. On its way to exit through the optic canal, the optic nerve passes medially to the ACP, which, as previously mentioned, extends superomedially to form the roof of the optic canal. The internal carotid artery (ICA) emerges from the distal dural ring along the inferomedial surface of the ACP. The oculomotor nerve courses inferolaterally to the ACP as it enters the roof of the cavernous sinus.

Al-Mefty has classified clinoidal meningiomas into three groups based on the site of origin and the adhesiveness of the tumor to the ICA. In group I, the tumor originates from the inferior surface of the ACP and adheres to the naked adventitia of the ICA prior to its investment by the arachnoid as it enters the subdural space from the cavernous sinus. This direct attachment of the tumor to the ICA without intervening arachnoid makes dissection of the tumor from the arterial wall impossible. Group II tumors originate from the superior and/or lateral surface of the ACP, away from the ICA as it emerges from the cavernous sinus. While these tumors may displace or encase the ICA and its branches with growth, an intact layer of arachnoid will separate the tumor from the adventitia of the vessels, making dissection feasible. Finally, group III tumors have their origin at the optic foramen and grow into the optic canal. These tumors typically become symptomatic early and are discovered when they are still small.

With its proximity to the optic nerve, clinoidal meningioma most commonly cause monocular visual disturbance. Optic canal involvement has been reported in 7 to 77% of clinoidal meningiomas. Visual outcomes following clinoidal meningioma resection have been mixed, with rates of postoperative visual improvements ranging from 10 to 84.6%. However, postoperative visual deterioration and complete visual loss have also been reported in multiple series ranging from 4 to 32%.

While total resection is the ideal goal of all meningioma operations, the surgical objectives for each case must be individualized. Surgical goals are designed by considering

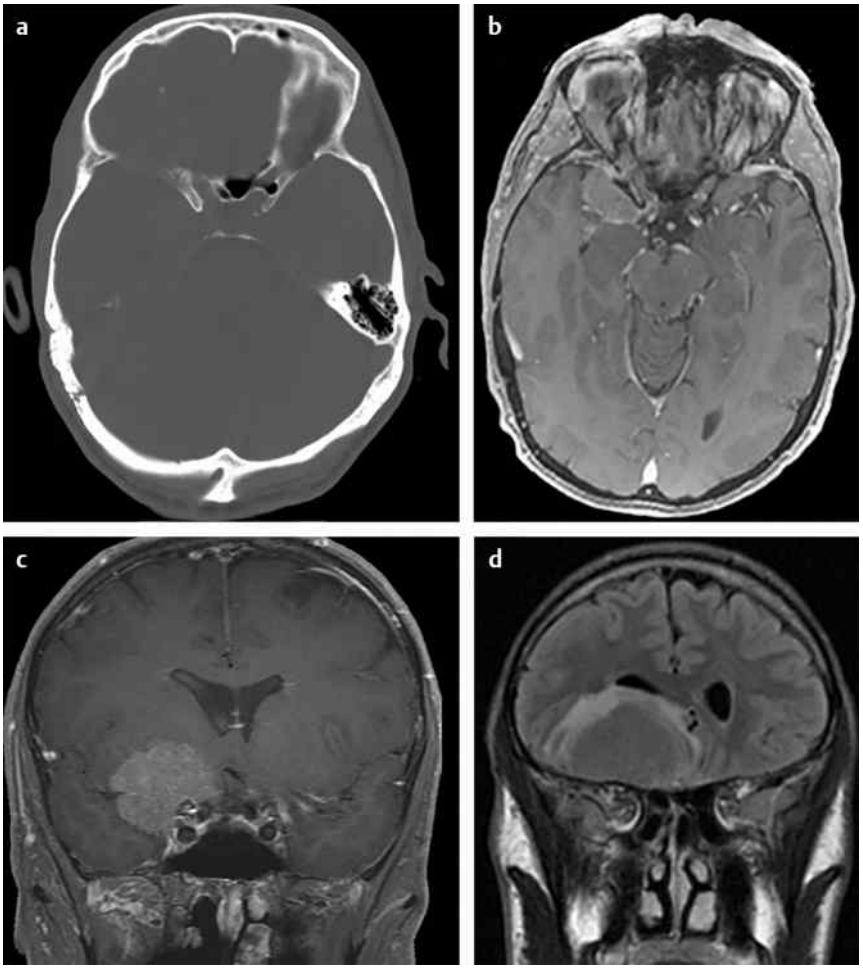


Fig. 4.1 Preoperative images. (a) CT showed that the right anterior clinoid process (ACP) was slightly hyperostotic. **(b)** MRI confirmed this finding; **(b)** and **(c)** showed an extra-axial lesion originating from the right ACP and **(d)** showed cerebral edema around this lesion.

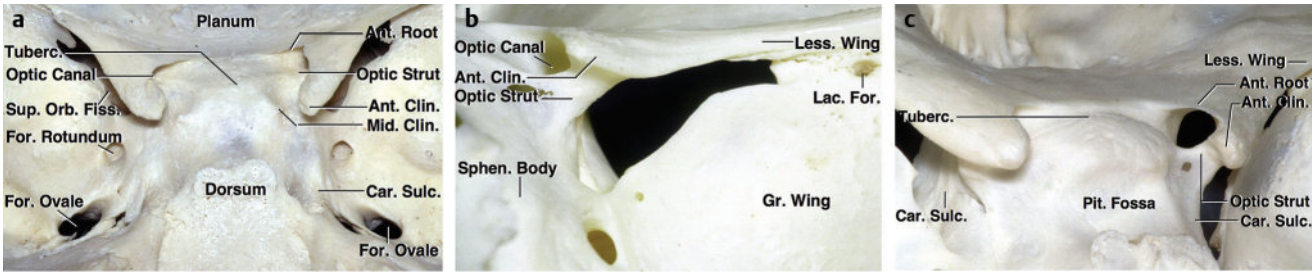


Fig. 4.2 Osseous relationships of the anterior clinoid process (ACP). (a) Superior view. The osseous structures, which nearly encircle the clinoid segment of the internal carotid artery, include the anterior clinoid laterally, the optic strut anteriorly, and the carotid sulcus medially. The anterior clinoid process projects backward from the lesser wing of the sphenoid bone, often overlapping the lateral edge of the carotid sulcus. The anterior root of the lesser sphenoid wing extends medially to form the roof of the optic canal. The posterior root of the lesser wing, referred to as the optic strut, extends from the inferomedial aspect of the anterior clinoid to the sphenoid body. The bony collar around the carotid artery, formed by the anterior clinoid, optic strut, and carotid sulcus, is inclined downward as it slopes medially from the upper surface of the anterior clinoid to the carotid sulcus. Another small prominence, the middle clinoid process, situated on the medial side of the carotid sulcus at the level of the tip of the anterior clinoid process, projects upward and laterally. In some cases, there is an osseous bridge extending from the tip of the middle clinoid to the tip of the anterior clinoid. In well-pneumatized sphenoid bones, the carotid sulcus is seen as a prominence in the lateral wall of the sphenoid sinus just below the floor of the sella. **(b)** Posterior view of the optic strut, optic canal, and superior orbital fissure. The optic strut separates the optic canal and superior orbital fissure and forms the floor of the optic canal and the superomedial part of the roof of the superior orbital fissure. The posterior surface of the strut is shaped to accommodate the anterior wall of the clinoid segment of the ICA. The artery courses along and may form a groove in the medial half of the lower aspect of the anterior clinoid before turning upward along the medial edge of the clinoid. **(c)** Oblique posterior view of the right optic strut. The lateral part of the bony collar around the clinoid segment is formed by the anterior clinoid, and the anterior part is formed by the posterior surface of the optic strut and the part of the carotid sulcus located medial to the anterior clinoid process. The posterior surface of the optic strut is wider medially adjacent to the carotid sulcus than it is laterally at the site of attachment to the anterior clinoid process. The optic strut slopes downward from its lateral end so that the medial part of the bony collar is located below the level of the part of the collar joining the anterior clinoid. (Reproduced from Laws E, Sheehan J, eds. Sellar and Parasellar Tumors. Diagnosis, Treatments, and Outcomes. 1st ed. Thieme; 2011.)

clinical and radiographic features of the tumor along with its expected natural history, and patient characteristics, such as age and medical comorbidities. The options of radiosurgery and radiotherapy for tumor control must also be deliberated. A thorough understanding of these factors, along with an appreciation for the patient's desires and wishes, will serve as the foundation on which to formulate a surgical strategy.

In addition to the basic tenets of meningioma surgery, such as early devascularization and early identification of neurovascular structures, a few specific technical factors must be considered when constructing a surgical approach to clinoidal meningioma. The craniotomy must be optimized for access to the tumor both for visualization and flexibility to manipulate instruments. The latter is frequently referred to as "surgical freedom." The bone opening must also be tailored and modified to minimize the need for brain retraction. If resection of the clinoid is indicated, then the advantages and disadvantages of extradural versus intradural clinoidectomy need to be contemplated. Finally, whether the tumor invades the optic canal or not, when the canal should be unroofed, whether routinely or only in selected cases, needs to be carefully considered preoperatively.

Options for Approach

Within the anterolateral corridor, pterional and more complex cranial base approaches have been advocated for the resection of clinoidal meningiomas. A pterional craniotomy provides transsylvian access to the tumor. As the sylvian fissure is split in a distal-to-proximal direction in this technique, the posterior capsule of the tumor is encountered first (**Fig. 4.3**). Identification of normal distal middle cerebral artery (MCA) branches provides the roadmap back to the proximal MCA, ICA bifurcation, the optic nerve and chiasm, and finally, the oculomotor nerve. This "traditional" craniotomy opening has the advantage of being "straightforward" and familiar to most surgeons. With this approach, the potential complications of more extensive cranial base osteotomies are not relevant. However, visualization and surgical freedom when attacking tumors with significant rostral extension is limited. Furthermore, with this intradural transsylvian method of tumor resection, early devascularization of tumor at the skull base is not possible. The identification of the ICA, the optic and oculomotor nerves happen late in the procedure, as it is possible only after a significant portion of the tumor has be removed.

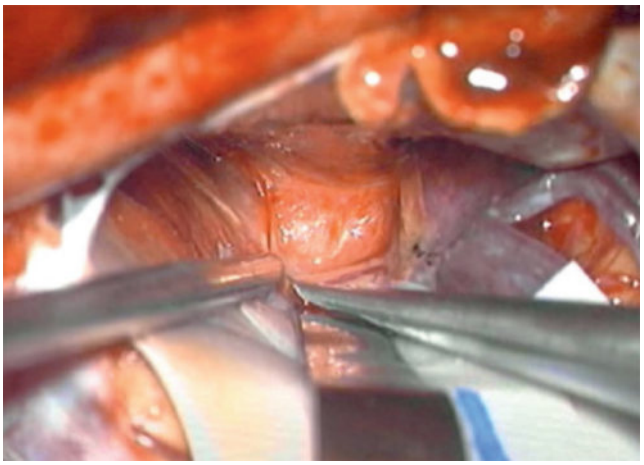


Fig. 4.3 Pterional approach. Intraoperative images show a clinoid meningioma at initial exposure after widely opening the sylvian fissure. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

The addition of a clinoidectomy permits early devascularization of the tumor and resection of tumor-infiltrated bone. This is particularly true for an extradural clinoidectomy, where the medial sphenoid wing and ACP may be aggressively resected while the intradural neurovascular structures are still protected under the dura. By removing the potentially hyperostotic ACP extradurally, the affected infrafrontal dura is exposed and the underlying tumor may be devascularized. However, the extradural technique is technically challenging and time consuming. Alternatively, the ACP can be removed after the dura is opened. This intradural method starts with exposure of the ACP under the frontal lobe, followed by incision of the dura overlying the ACP, often in the shape of a "T" to expose the bony ACP. Thereafter, this is drilled away with either a diamond burr or a similar tool. This technique allows a more selective and tailored removal of the ACP, but the intradural neurovascular structures are fully exposed and potentially vulnerable to injury from the drill. Bone dust from the drilling can disperse in the subarachnoid space and potentially cause meningeal irritation. Furthermore, the tumor itself may hinder the intradural exposure of the ACP and surrounding structures, and therefore, for large tumors, intradural anterior clinoidectomy may have to wait until after significant bulk of the tumor has been removed.

When a clinoidectomy is considered, either intradurally or extradurally, the preoperative CT must be studied carefully for potential anatomical variants. The presence of a pneumatized ACP represents an increased risk for postoperative cerebrospinal fluid (CSF) leak following a clinoidectomy, and extra care must be taken to repair the dural defect. An osseous protuberance may connect the tip of the anterior clinoid to the middle clinoid to form a bony ring around the ICA called the caroticoclinoidal foramen. There may also be an interclinoid osseous bridge between the anterior and the posterior clinoid processes. These bony connections between the anterior clinoid with the middle and posterior clinoid create additional challenges to removal of the anterior clinoid.

The routine unroofing of the optic canal during clinoidal meningioma surgery remains controversial. The procedure carries risks for either direct or thermal injury to the optic nerve. However, unroofing of the optic canal, especially performed as part of an extradural clinoidectomy, along with sectioning of the falciform ligament over the optic nerve, provides early decompression and mobilization of the optic nerve. This relieves the compression from tumor infiltration into the optic canal and furthermore, prevents traction injuries to the optic nerve during subsequent tumor manipulations. In one case series, a trend for improved visual outcomes was seen for patients who underwent unroofing of the optic canal.

Approach of Choice

A thorough understanding of these anatomical nuances and surgical techniques is necessary to design a surgical approach. In this case, the decision was made to proceed with a one-piece modified right orbitozygomatic craniotomy with extradural clinoidectomy. The addition of a supraorbital osteotomy to the frontotemporal

craniotomy will expand the surgical corridor to the superior pole of this large tumor. The extradural clinoidectomy with unroofing of the optic canal will provide early decompression of the optic nerve, tumor devascularization, and identification of the critical neurovascular structures at the skull base.

Questions

1. If the supraorbital osteotomy violated the frontal sinus, how would you handle the sinus to prevent CSF rhinorrhea?
2. What precautions can you take to avoid injury to the frontalis branch of the facial nerve while developing the scalp flap for this craniotomy?

■ Description of the Technique

The patient was positioned supine with chest elevated 10 degrees and the head rigidly fixated in the Mayfield three-pin head holder. The head was rotated 30 degrees to the left and tilted toward the floor such that the malar eminence was brought to the highest point of the operative field. The patient's facial features were registered to previously obtained MRI scan for stereotactic navigation. Mannitol, Decadron, and mild hyperventilation were utilized for brain relaxation (see **The Three Approach Elements).**

The Three Approach Elements

Corridor: anterolateral

Craniotomy: "pterional"

Modifiers: supraorbital osteotomy, extradural clinoidectomy

A curvilinear incision was made behind the hairline starting 1 cm anterior to the tragus, adjacent to the root of the zygoma, and ending just past the midline. The scalp was elevated with the pericranium left on the cranium. The superficial temporal fascia was incised from the root of the zygoma to the keyhole. Dissection was carried through the fat pad down to the deep temporal fascia, and an interfascial dissection was performed to avoid injury to the frontalis branch of the facial nerve. This dissection continued until the superior edge of the zygomatic arch and the lateral orbital rim were exposed. The pericranium was cut laterally along the superior temporal line and posteriorly along the skin incision. It was carefully elevated off of the cranium and preserved. The superior orbital rim was exposed medially to the supraorbital notch. The periorbita was dissected free from the inner surface of the orbit from the supraorbital notch to just past the frontozygomatic suture laterally. The temporalis fascia was cut along the superior temporal line while preserving a 1 cm myofascial cuff, and along the posterior edge of the scalp incision. The temporalis muscle was elevated and reflected inferiorly, exposing the pterion (see **Operative Setup**).

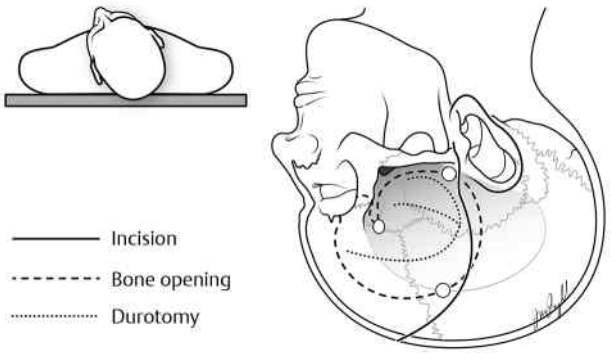
Operative Setup

Position: supine, head turned 30 degrees to the left

Incision: curvilinear incision

Bone Opening: one-piece modified orbitozygomatic craniotomy

Durotomy: C-shaped with linear cut over the sylvian fissure toward the optic canal



Craniotomy

A one-piece modified orbitozygomatic craniotomy (fronto-temporal craniotomy with supraorbital osteotomy) was performed using standard techniques. First, a MacCarty burr hole was placed over the frontosphenoidal suture approximately 1 cm posterior to the frontozygomatic junction (**Fig. 4.4**). The frontal dura was exposed in the superior portion of the burr hole, the periorbita was exposed in the inferior portion, and the orbital roof was seen separating the two halves. A temporal burr hole was placed slightly anterior and superior to the root of the zygoma. Using the craniotome, a cut was made from the temporal burr hole extending superiorly and anteriorly to the supraorbital ridge, just lateral to the supraorbital notch (**Fig. 4.5**). Starting at the frontal portion of the MacCarty burr hole, a cut was made inferiorly and posteriorly across the sphenoid ridge to reach the temporal burr hole. Continuing with the craniotome, a cut was made from the orbital portion of the MacCarty burr hole travelling inferiorly along the lateral orbital wall toward the inferior orbital fissure and stopping just beyond the frontozygomatic suture. At this point, the drill was turned outward 90 degrees and brought across the lateral orbital wall. The initial bone cut that terminated at the superior orbital ridge was extended across the orbital rim and 1 cm into the orbital roof using a combination of drill and osteotome. Finally, a small osteotome was used to cut across the orbital roof from the MacCarty burr hole to the medial orbital bone cut. Once the one-piece cranio-orbital bone flap was elevated, additional bone from the anterolateral portion of the orbital roof was removed and the lesser sphenoid wing was resected down to the superior orbital fissure and meningo-orbital band.

Extradural Clinoidectomy

At this point, the microscope was brought into the operating field. The meningo-orbital band was cut approximately 5 mm (**Fig. 4.6a**), which allowed for further retraction of the frontotemporal basal dura (**Fig. 4.7a**). The ACP was exposed and appeared to be hyperostotic. Using a diamond drill with continuous irrigation, the orbital roof was thinned and removed with a fine curette. The optic nerve was identified within the optic canal (**Fig. 4.7b**). The central cancellous bone of the ACP was excavated and the optic strut was disconnected with the diamond drill. The remaining bone fragment was dissected free from the ligamentous attachment, removed with a pituitary rongeur and sent to pathology (**Fig. 4.7c**). Venous bleeding from the cavernous sinus was controlled with Fibrillar and Gelfoam.

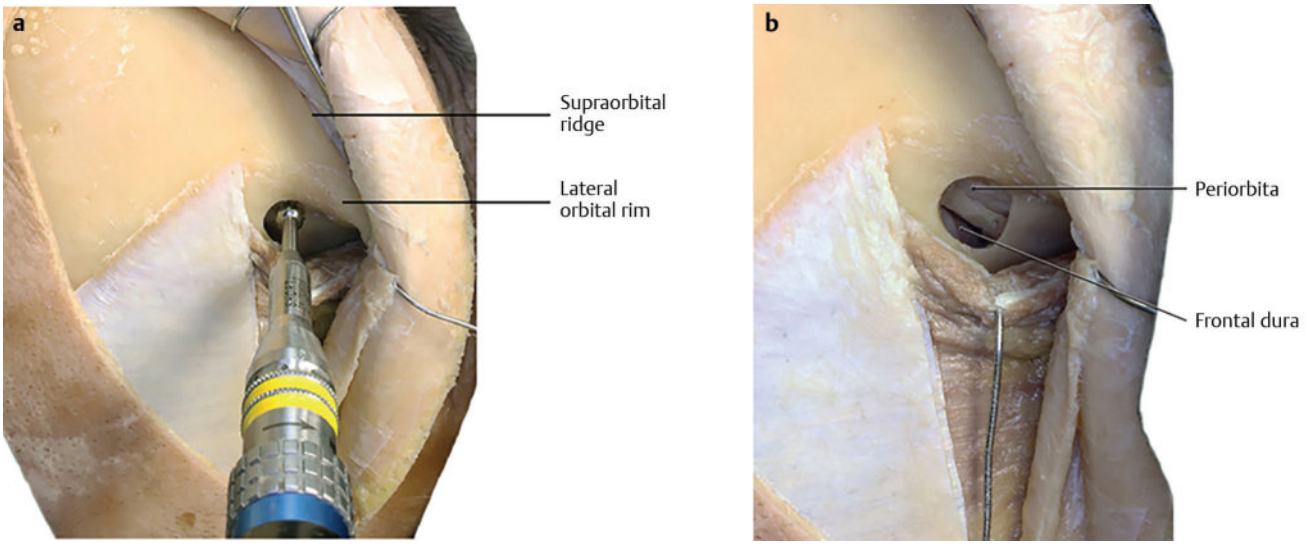


Fig. 4.4 Cadaveric dissection (right side) showing the placement of the MacCarty burr hole to access the anterior fossa and orbit. (a) The burr hole was placed over the frontosphenoidal suture approximately 1 cm posterior to the frontozygomatic junction. **(b)** The frontal dura was exposed in the superior portion of the burr hole, the periorbita was exposed in the inferior portion, and the orbital roof was seen separating the two halves. (Reproduced from Spetzler R, et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

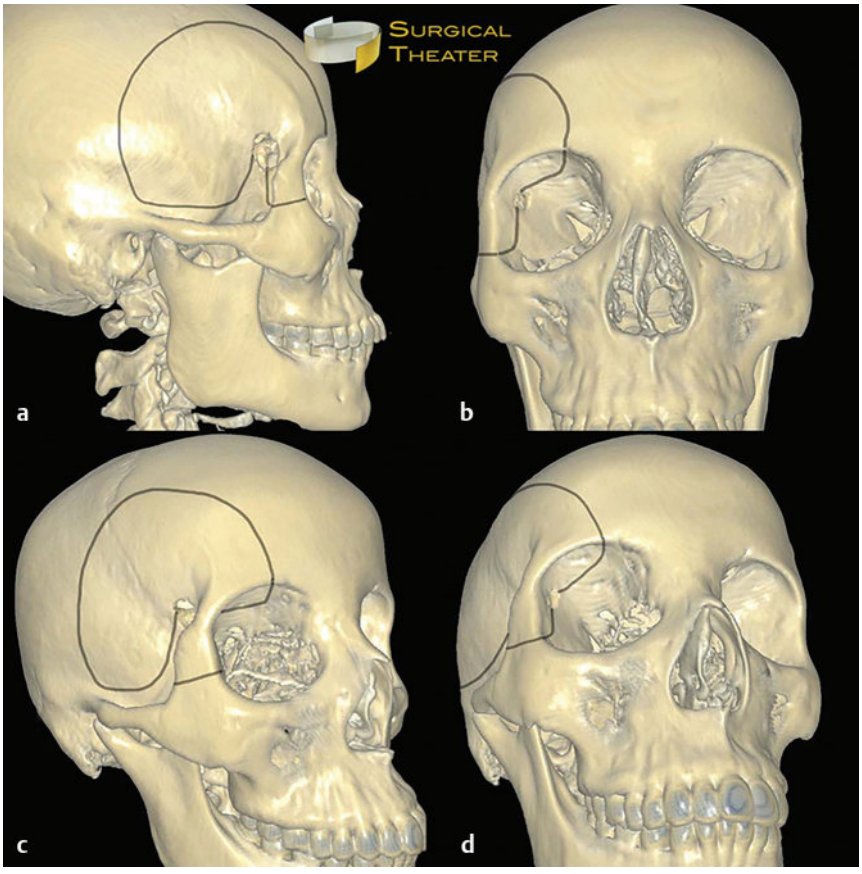


Fig. 4.5 3D rendering of the skull and the one-piece, modified orbitozygomatic craniotomy (with supraorbital osteotomy). (a–d) Lateral, AP, and various oblique views of the skull and the one-piece, modified orbitozygomatic craniotomy (with supraorbital osteotomy). The lines demonstrate the bone cuts of the technique. Note that in this case, a temporal burr hole was not used. From the frontal portion of the MacCarty burr hole, the craniotome is run in a nearly complete circle, ending just lateral to the supraorbital notch. It is then used to make the lateral orbital cut, starting in the orbital portion of the MacCarty hole, moving inferiorly in the direction of the inferior orbital fissure, and then turned 90 degrees to cut across the lateral orbital wall. The orbital roof cuts are then completed with osteotomes.

Tumor Resection

The dura was opened in a curvilinear fashion. Upon elevation of the dura, the tumor was easily identifiable in the proximal sylvian fissure. The dissection of the tumor was first focused on defining the capsule and margins of the tumor. The sylvian fissure was opened widely in a distal-to-proximal direction by following M2 branches. The tumor was noted to be very firm, well-encapsulated, and quite fibrous. Next, the dura was split along the sylvian fissure toward the optic nerve in the optic canal. The falciform ligament over the optic nerve was cut for decompression. Tracing the optic nerve back from its extradural component, the cisternal segment was identified within the tumor. The arachnoid was quite thickened over the optic nerve,

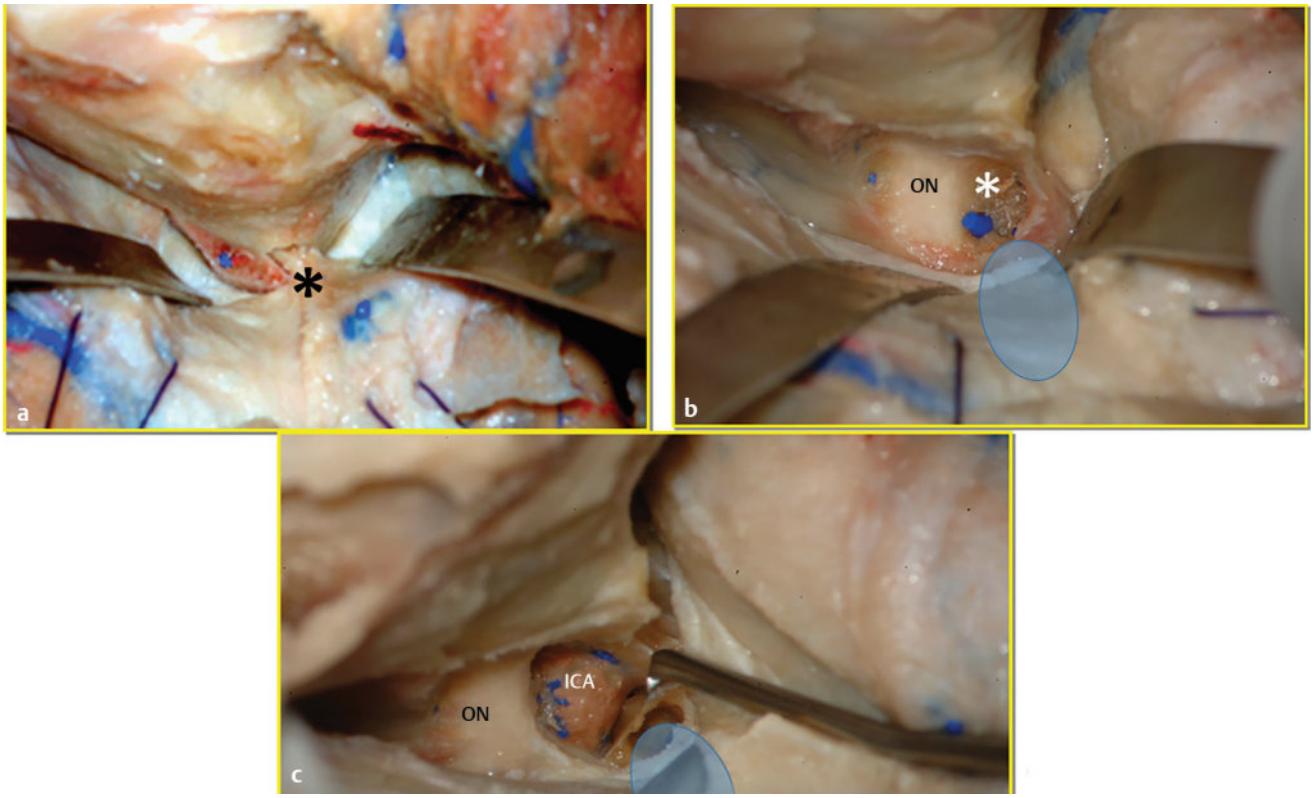


Fig. 4.6 Cadaveric dissection showing an extradural clinoidectomy. (a) The lesser wing of the sphenoid was drilled away until the superior orbital fissure was unroofed. The meningo-orbital band (*black asterisk*) is shown. This is subsequently cut to allow further retraction of the frontotemporal basal dura. **(b)** The roof of the orbit was removed with a diamond burr until the optic nerve was uncovered. The optic strut (*white asterisk*) would be removed next. The *blue oval* denotes the position of the anterior clinoid process (ACP) below the dura. **(c)** After drilling the optic strut, the ACP was outfractured and removed. Note that in this specimen, the ACP was pneumatized. ON, optic nerve; ICA, internal carotid artery.

but a dissection plane was nevertheless developed between the tumor and optic nerve. Dissection was carried along the anterior skull base in order to devascularize the tumor. The tumor was particularly adherent to the portion of the dura that covered the ACP, optic canal, and the planum. The affected dura was removed along with the tumor.

After the tumor had been detached from the anterior skull base, attention was shifted to identification of critical neurovascular structures. Returning to the exposed right optic nerve at the skull base, tumor over the nerve was removed and the cleavage plane between the tumor and the optic nerve was carried toward and then across the optic chiasm to the left optic nerve (**Fig. 4.7d**). No tumor was noted in the prechiasmatic cistern. Working lateral to the right optic nerve, the right ICA was identified, and the adventitia was noted to be free from tumor invasion. At this point, the bulk of the remaining tumor limited further dissection. Therefore, the tumor beneath the frontal lobe was aggressively debulked. As the tumor was removed, the tumor capsule was mobilized into the working space and coagulated and cut. Initially, there was a good dissection plane between the tumor capsule and the parenchyma, but the tumor became more adherent to the brain as the resection continued more superiorly into the frontal lobe. Given the T2 signal abnormalities on the preoperative MRI, this was the anticipated finding (**Fig. 4.1d**). With the frontal portion of the tumor removed, the M2 branches were dissected free from the tumor capsule and traced back to M1. The tumor overlying M1 was much more adherent to the adventitia and to the perforating vessels arising from its dorsal surface. To avoid a vascular injury, a small remnant of tumor was left attached to the M1 segment and to its perforators. The final remaining tumor in the middle cranial fossa was removed and the cisternal portion of the right oculomotor nerve was visualized on its way to the oculomotor triangle. There was no obvious cavernous sinus involvement.

For closure, the dura was closed in a watertight fashion with a dural patch using the previously harvested pericranium. The anterior clinoidectomy site was packed with Fibrillar and Gelfoam. The frontal sinus was entered during the craniotomy, Gelfoam with iodine was packed into the sinus and a piece of pericranium was rotated with a pedicle to cover the defect. A central tack-up was placed and the bone flap was secured back into place using titanium plates and screws. A Medpor pterional graft was placed over the pterion for cosmesis and support. The temporalis muscle and the scalp were closed in layers.

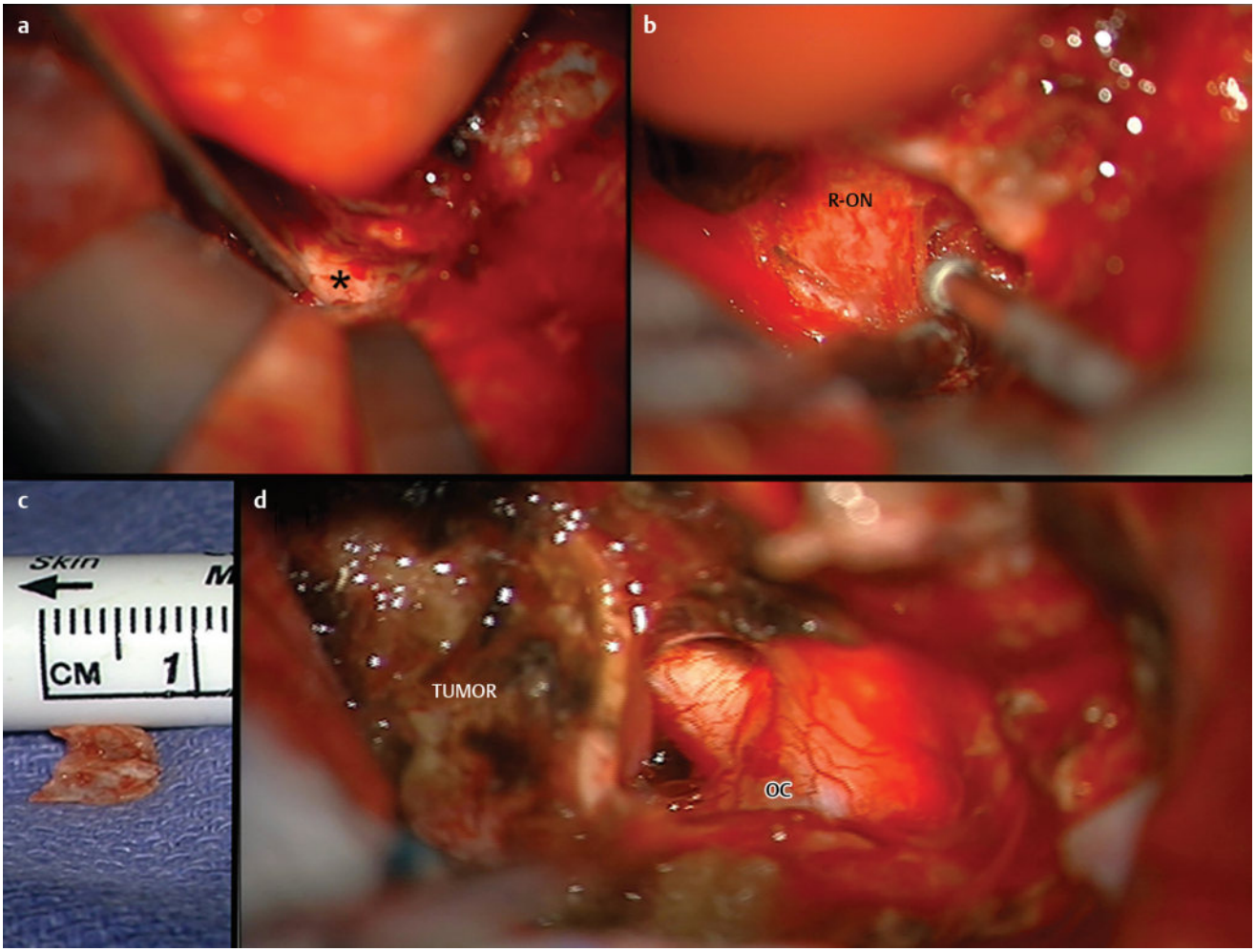


Fig. 4.7 Intraoperative images. (a) After cutting the meningo-orbital band, the anterior clinoid process (ACP shown with*) was exposed. **(b)** The optic strut was removed with a diamond burr. R-ON, right optic nerve. **(c)** The ACP was removed extradurally. **(d)** Under the microscope, the tumor was dissected off of the optic apparatus. OC, optic chiasm.

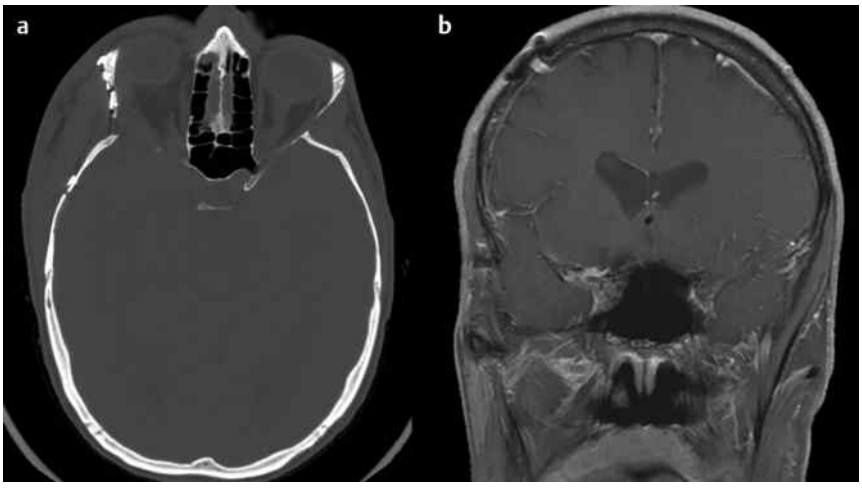


Fig. 4.8 Postoperative images. (a) CT showed the removal of the right ACP. **(b)** Except for a small residual, left intentionally to avoid injury to the MCA, the tumor was removed successfully.

Surgical Pearls

1. While raising the skin flap off the frontal bone, it is important to preserve the pericranium. If the craniotomy violates the lateral corner of the frontal sinus, this can be used to as a vascularized flap to cover this area to prevent postoperative CSF rhinorrhea. Surgeons "harvest" the pericranium in different ways; some like to keep it with the galea initially and separate it from the skin/galea side while others like to keep it with the bone initially and raise it from the bone before use. It is our preference to do the latter, as it seems to yield more robust and intact tissue for the use of closing the sinus.
2. A relaxed brain can greatly facilitate the extradural clinoidectomy. As such, a lumbar drain, and in the appropriate setting, an external ventricular drain, can make the clinoidectomy easier and safer. If these are somehow not feasible, a small durotomy in the frontal basal dura can provide access for CSF aspiration. This should be done early in the procedure, right after the elevation of the bone flap, to allow enough time for CSF egress before the clinoidectomy.
3. The release of the meningo-orbital band (**Fig. 4.6a**) is critical for gaining sufficient exposure to perform the extradural clinoidectomy. If the incision follows the contour of the temporal lobe dura, as it curves from anterolateral to posteromedial, there is minimal danger that the oculomotor nerve would be injured during the process.
4. Preoperative evaluation of the thin-cut CT is critical to detect any anatomical variant related to the ACP. Some variants, such as the osseous bridges mentioned above, can make the clinoidectomy impossible. Others, such as pneumatization of the clinoid (**Fig. 4.6**), can increase the chance of postoperative complications.

■ Aftercare

The patient had a routine postoperative hospital course with no new neurological deficits. The blurry vision he experienced preoperatively improved after surgery. The images taken after his operation confirmed that the tumor, as well as the right ACP, had been successfully removed (**Fig. 4.8**). It also showed a very small remnant of the tumor that was left intentionally to avoid injury to the MCA. The portion of the tumor has been stable for 3 years, during subsequent follow-up of the patient.

■ Possible Complications and Associated Management

Aside from routine steroid and anticonvulsants to prevent postoperative cerebral edema and seizures, respectively, evaluation of the patient's vision and oculomotility should be performed as soon as the patient wakes from anesthesia. The right eye will swell shut, usually between the first and third postoperative day, and this will make evaluation of the ipsilateral eye very difficult if not impossible. The operation was performed in close proximity to the optic apparatus as well as the oculomotor nerve, and therefore, these vision examinations should be done as thoroughly as possible.

Despite great care before and during surgery, small openings into the frontal sinus (during the craniotomy) and ethmoid sinus (during the clinoidectomy) can sometimes go unnoticed. Patients should be monitored for postoperative CSF rhinorrhea.

Perspective

Nikolai J. Hopf

■ Introduction

Anterior clinoid meningiomas are defined by their origin from the area of the ACP, but these tumors have a widely diverse growth pattern. Generally, all of them grow to some extent laterally along the sphenoid ridge. A subgroup of tumors extends predominantly superiorly along the planum, anterior skull base, and suprasellar region. Others show a significant inferior extension along the lateral wall of the cavernous sinus and/or the lesser sphenoid wing.

The most frequently used surgical route for anterior clinoid meningiomas is the transsylvian corridor because it allows adequate exposure of both the superior and inferior extension of these tumors. To access the transsylvian corridor, one can consider the standard pterional craniotomy, with or without the orbitozygomatic extension, as well as the minimally invasive variant through a minipterional keyhole approach. Alternatively, anterior clinoid meningiomas that extend predominantly superiorly and anteriorly may be resected using a subfrontal corridor. This can be accessed via a standard frontolateral craniotomy using a frontolateral skin incision in the hairline or a bicoronal incision. A minimally invasive variant of this, the supraorbital keyhole craniotomy using an incision within the eyebrow, is also worth considering (**Fig. 4.9**). Furthermore, recent reports have suggested that the endonasal endoscopic approach can be utilized for selected anterior clinoid meningiomas.

The transsylvian corridor has obvious advantages as it provides access to both the inferior (temporal) and superior (frontal) parts of tumors around the ACP. However, it has several disadvantages as well. Maximal exposure requires extensive soft tissue manipulation, particularly around the temporalis muscle, which can lead to postoperative discomfort and poor cosmetic results. The approaches, such as the one described in the first section of this chapter, are time consuming and associated with additional risks for the intraorbital structures and the temporomandibular joint.

Endoscopic keyhole approaches are designed to overcome these disadvantages. The chief tenet for the keyhole concept is that, through strategically placed small openings, surgical corridors are accessed with sufficient but not excessive space to maneuver instruments, as the endoscope brings illumination up to the target, thus minimizing iatrogenic trauma to the patient from incision to resection (**Fig. 4.10**). Reisch et al have shown that the supraorbital craniotomy using an eyebrow incision is associated with minimal postoperative discomfort and cosmetic changes. The downside of keyhole craniotomies is that they provide only a single, narrow trajectory to the lesion and a strictly parallel view along the surgical instruments. As such, skilled handling of endoscopes to generate diverse viewing angles is required to adequately visualize the lesion and its surroundings, and delicate surgical technique is necessary to maneuver instruments in the narrow corridor. Generally, it can be said that extensive approaches are more convenient for the surgeon, keyhole approaches for the patient.

■ Case Presentation

A 79-year-old man presented with a history of progressive listlessness, memory deficit, and tiredness for the last few months. On examination, he was alert but only partially oriented. He was able to recall two out of three objects at 1 minute. His cranial nerve, motor, and sensory examinations were unremarkable. His MRI revealed a well-demarcated and contrast-enhancing tumor, originating from the ACP on the right side with significant peritumoral edema (**Fig. 4.11**). The tumor measured 4.7 cm in the mediolateral, 4.0 cm in the cranial–caudal, and 4.7 cm in the anterior–posterior dimensions. A meningioma was the suspected diagnosis.

■ Anatomical and Therapeutic Considerations

In addition to an extension along the sphenoid ridge, this tumor showed a predominantly superior/anterior growth pattern, and significant peritumoral edema in the frontal and temporal

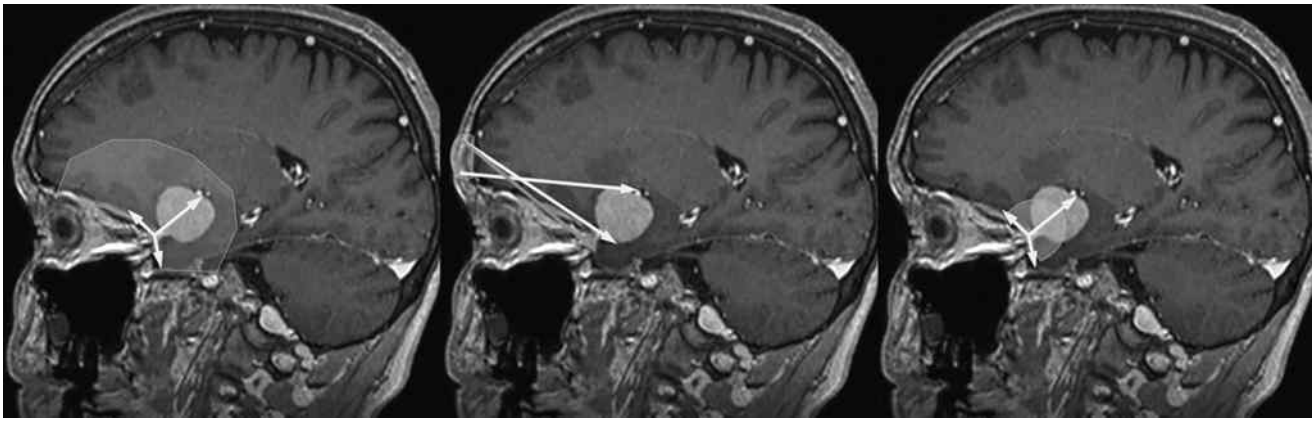


Fig. 4.9 Various approaches to an anterior clinoid meningioma. Sagittal T1-weighted MRI with contrast of a patient with an anterior clinoid meningioma depicting the craniotomy and possible dissection direction of a standard pterional (*left*), supraorbital keyhole (*middle*), and minipterional keyhole craniotomy (*right*).

lobes. Even though radical resection is generally the surgical goal in meningiomas, gross total resection was not the highest priority in this elderly patient with only minor complaints. Surgical goals in this particular case were defined as reduction of the mass effect, decompression of the brain, optic nerves and blood vessels, as well as reduction of the edema with minimal approach-related trauma.

Because of the particular growth pattern of this tumor, the supraorbital approach through the subfrontal corridor is highly suitable for accomplishing all the above-mentioned goals. In addition, it offers the possibility of achieving a gross total resection. To minimize the iatrogenic, approach-related injuries in this elderly patient, the keyhole craniotomy was chosen. Despite the large size of the tumor, it involved predominantly the anterior cranial fossa. Therefore, the subfrontal route provides the opportunity for tumor devascularization and identification of the critical neurovascular structures early in the procedure, as well as adequate access to all parts of the tumor.

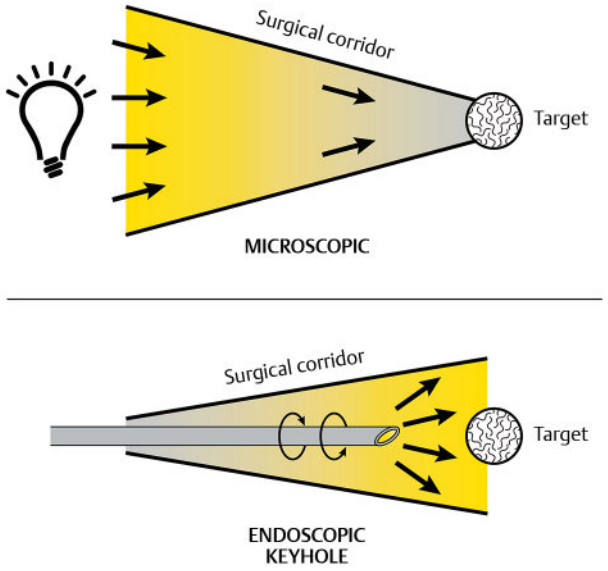


Fig. 4.10 Illustration of the "endoscopic keyhole" concept. For "open" microsurgery, illumination of the target is dependent on the light source on the microscope. Because of this, the opening to the corridor must be sufficiently wide to "bring in the light," leading to the doctrine that "the incision must be bigger than the craniotomy; the craniotomy must be bigger than the tumor." Under the keyhole concept, the endoscope brings light close to the target through strategically placed small openings, making it sometimes possible that the corridor may in fact widens as it deepens. At depth, rotation of an angled endoscope can provide various viewing angles of the tumor and surroundings.

Description of the Technique

The patient was positioned supine, similarly as described earlier for the pterional approach. The patient's chest was slightly elevated, and the head rigidly fixed in the Mayfield head holder turned 30 degrees to the left side. The neck was slightly extended. Navigation, mannitol, Decadron, and hyperventilation were not utilized.

A 4-cm skin incision was made within the lateral aspect of the right eyebrow, not extending into the muscle layer. The skin was then elevated from the fascia of the frontalis muscle and from the superficial fascia of the temporal muscle, but only along the most anterior aspect of the superior temporal line. Then, the frontalis muscle was incised approximately 1 cm superior to its attachment with the orbicularis oculi muscle in a medial-to-lateral direction, between the supraorbital nerve medially and the superior temporal line laterally. The temporal fascia was then detached from the superior temporal line. Hooks were used to keep the soft tissue apart.

A burr hole was performed immediately lateral to the superior temporal line and enlarged with a Kerrison punch toward the floor of the anterior skull base. Using the craniotome, a 2-cm long cut was made parallel to the frontal skull base in a lateral-to-medial direction. Then, the craniotome was brought back to the burr hole and the craniotomy completed in a C-shaped manner superiorly in a lateral-to-medial direction. With that, a 2 × 1.5 cm fronto-latero-basal craniotomy was achieved (**Fig. 4.12a**). The dura was bluntly dissected off the frontal skull base enabling removal of the inner edge of the bone and a couple of juga, or ridges of the orbital roof using a diamond drill.

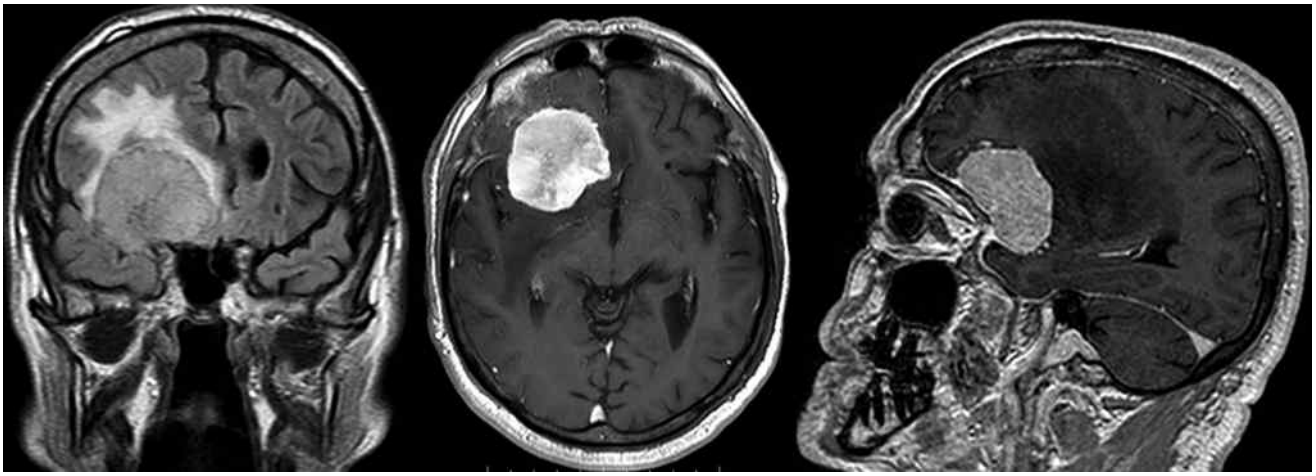


Fig. 4.11 Preoperative images. A 79-year-old male patient with a large anterior clinoid meningioma with a coronal FLAIR image demonstrated the superior-to-inferior extension and edema of the frontal lobe (*left*). T1-weighted images with contrast showed the tumor extension in anterior-to-posterior and medial-to-lateral direction (*middle*), as well as the predominant superior and anterior growth pattern of the tumor (*right*).

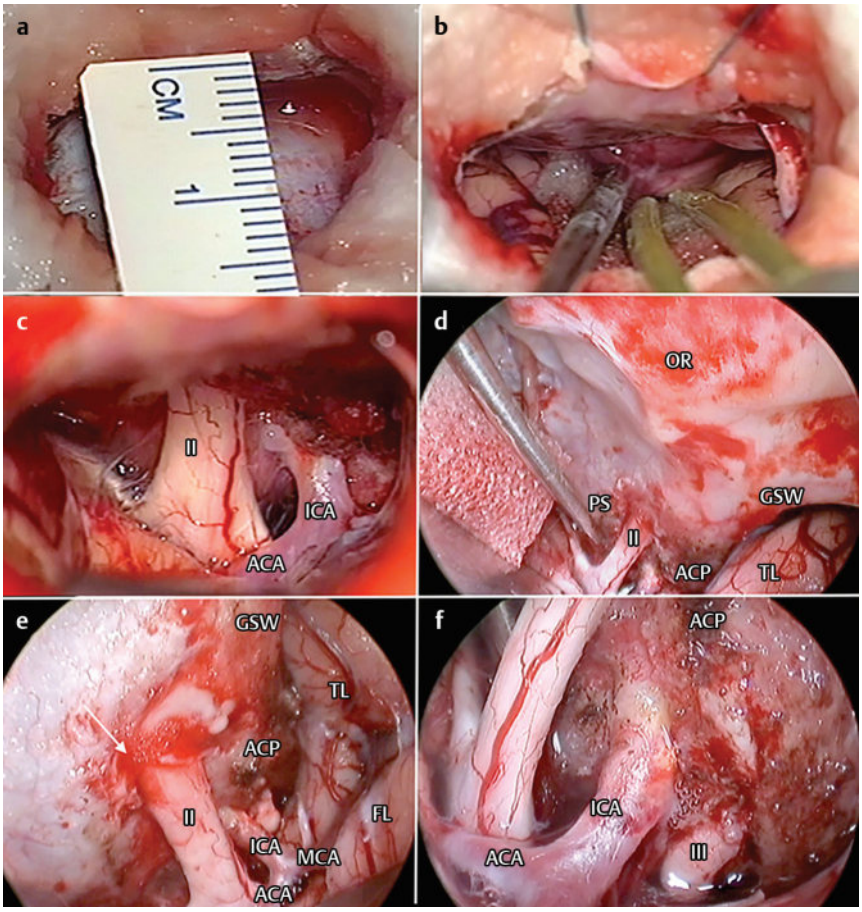


Fig. 4.12 Intraoperative views. **(a)** Microscopic view before opening of the dura; **(b)** microscopic view during an effort to open the basal cisterns for CSF release and **(c)** after partial removal of the tumor giving view to the optic nerve (II), internal carotid artery (ICA), and anterior cerebral artery (ACA). **(d)** Endoscopic images of the same patient after gross total tumor resection providing a wide overview of the entire region including the orbital roof (OR), greater sphenoid wing (GSW), planum sphenoidale (PS), anterior clinoid process (ACP), optic nerve (II), and temporal lobe (TL). **(e)** Close-up looking 30 degrees laterally demonstrating the opened falciform ligament (*arrow*) to release the optic nerve and the middle cerebral artery (MCA) running toward the sylvian fissure between the intact frontal lobe (FL) and temporal lobe. **(f)** Close-up looking 30 degrees medially demonstrating the coagulated dural base of the tumor around the anterior clinoid process (ACP) and internal carotid artery (ICA) as well as the preserved oculomotor nerve (III).

The dura was opened in a curvilinear fashion and reflected inferiorly. The frontal lobe was appearing full but pulsating slightly. It did not herniate transdurally despite the large size of the tumor and frontal lobe edema. The first effort to gain excess to the basal cisterns in order to release CSF was unsuccessful both lateral and medial to the tumor. Therefore, the tumor attachment at the base of the frontal lobe was attacked first with bipolar coagulation, in order to devascularize the tumor. Then, the anterior part of the tumor was decompressed. This was quite difficult because of the firm consistency and significant calcifications within the tumor.

After some debulking, the arachnoid to the basal cisterns and sylvian fissure was finally accessible, and it was opened with a diamond knife (**Fig. 4.12b**). CSF was slowly removed over 2 to 3 minutes. This led to a significant decrease of the intracranial pressure, allowing the frontal lobe to sink below the level of the dura by gravity, minimizing the need for retractors throughout the entire operation.

From this point on, the delicate dissection around the tumor was performed with bimanual technique, and the endoscope was held variably by the assistant, or, when a perfectly steady image was needed, by the endoscope holder. The tumor was initially found to be well-encapsulated and easily separable from the frontal and temporal lobe arachnoid. However, the tumor became more adherent to the brain as the resection continued, as predicted from the T2-signal abnormalities on the preoperative MRI. Step by step, the branches of the MCA were identified and dissected off the tumor capsule from distal to proximal. Then, the most medial aspect of the tumor, next to the optic nerve and carotid artery, was attacked. Opening the falciform ligament was performed to untether the right optic nerve and decrease the risk of iatrogenic nerve injury from operative manipulation. Keeping the arachnoid layers intact during the dissection around the nerve further facilitated this process. After that, the carotid artery, anterior cerebral artery, and most proximal aspect of the MCA were identified and dissected free (**Fig. 4.12c-e**). Fortunately, only a few tumor feeders from the cerebral arteries were present. A 30-degree angled endoscope was used at this point for inspection of the entire operative area, confirming complete tumor resection (**Fig. 4.12f**). Finally, vigorous bipolar coagulation of the dural base of the tumor was performed while meticulously avoiding the optic nerve.

For closure, the dura was reapproximated with standard sutures and watertight sealing achieved using TachoSil. The small bone flap was fixed to the skull with plates and screw, and the muscles and skin were closed in layers.

■ Aftercare

The patient had an uneventful postoperative course with no new neurological deficits. The early postoperative MRI confirmed gross total tumor removal and no complications (**Fig. 4.13**). The wound healed well with an excellent cosmetic result. The patient went quickly back to normal life and physical activities, and started to play tennis 4 weeks after surgery.

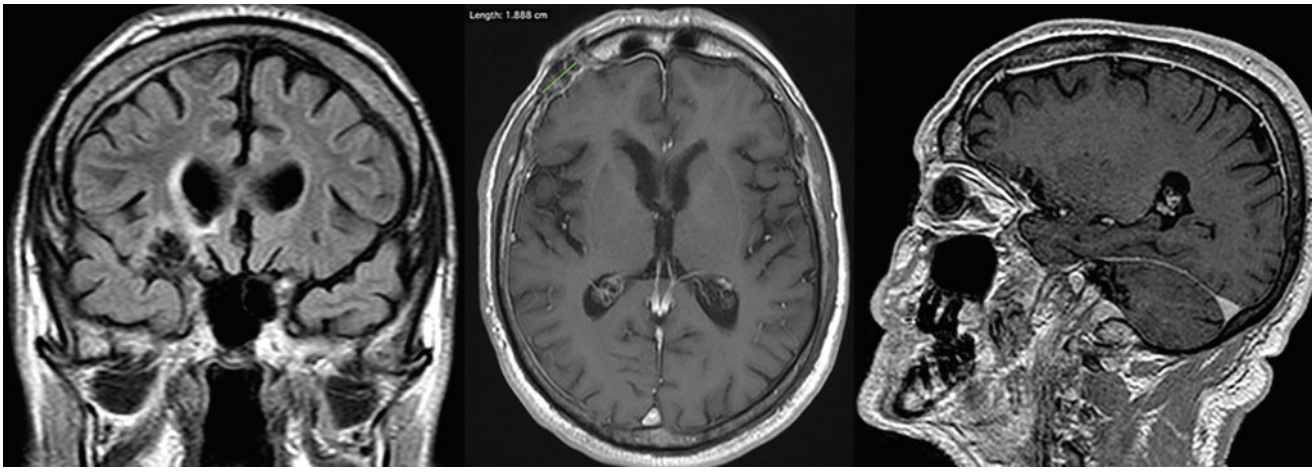


Fig. 4.13 Postoperative images. MR images of the same patient demonstrating gross total tumor resection and resolution of the edema 8 weeks after surgery in a comparable FLAIR image (*left*) and T1-weighted axial (*middle*) and sagittal (*right*) images with contrast through a right-sided supraorbital keyhole craniotomy of a size less than 2 cm (*middle*).

Fig. 4.14 Minipterional keyhole craniotomy. The clinoid meningioma presented in the first section has a component (*arrow*) which extended along the sphenoid wing into the anterior middle fossa. This portion would not be accessible through a supraorbital keyhole approach. A minipterional keyhole approach, however, may be useful. For this, the skin incision is a slightly curved "C," slightly behind the temporal hairline. A line drawn perpendicularly backward from the sphenoid wing should roughly bisect this "C." The head is positioned so that the malar eminence is the highest point in the field. The bone flap is hidden under the temporalis muscle and once elevated, the sphenoid wing is drilled flat as with a conventional pterional craniotomy.

■ Commentary

The case described in this section highlights the importance of minimally invasive treatment strategies in elderly patients. Minimally invasive technique in this context does not only mean the use of particularly small craniotomies. Rather, it should be understood as a concept for providing a highly tailored treatment strategy for the individual needs of a patient. In this case example, the primary goal was to reduce the mass effect of the tumor, with minimal iatrogenic injury to the elderly patient who had but a few symptoms. The goal was not a radical tumor resection.

An endoscopic keyhole approach provided sufficient visualization of this tumor with predominantly lateral, superior, and anterior extensions. The exposure was not only adequate to control all the critical structures nearby, but made complete tumor removal possible. The patient experienced a very favorable postoperative course and a *restitutio ad integrum* within a few weeks.

Limited openings also have significant disadvantages. Trajectory to the lesion is restricted to a single track, and both instruments and technique must be adapted to this. Furthermore, appropriate experience with handling endoscopes in narrow corridors is a prerequisite for adequate visualization of the target and successful application of the keyhole concept (**Fig. 4.10**).

The supraorbital keyhole craniotomy utilized in this case is only suitable for tumors without significant temporal extension. For tumors with an extension along the lesser sphenoid wing, in the anterior part of the middle fossa, and/or the lateral wall of the cavernous sinus, the transsylvian route must be used. The tumor presented in the first part of this chapter is such an example. However, a transsylvian route can also be realized using a keyhole craniotomy. The minipterional keyhole craniotomy reduces the "standard" pterional opening to only those parts which are absolutely necessary. This includes a small area of around 1.5 cm inferior, superior, and posterior to the pterion, providing a limited but very versatile craniotomy of around 2 × 3 cm (**Fig. 4.14**). This keyhole craniotomy requires a much smaller skin incision, resulting in less manipulation of

the temporal muscle and soft tissue. Apart from the size, the dissection direction and anatomical orientation are identical to the standard approach, keeping the surgeon in his/her " comfort zone."

Conversion from standard approaches to their keyhole variants is best achieved in a stepwise manner. This starts with analyzing, at the end of any operation, which part of the craniotomy opening was actually used versus that which was excess. This analysis will most likely lead to the conclusion that the exact same procedure can be done using a much smaller opening. A judicious, stepwise reduction of the size of the craniotomy and skin incision should be the next step in this evolutionary process. However, keyhole approaches, such as the supraorbital technique demonstrated here, involve not just smaller openings, but often also involve strategic relocation of the openings, which can transform the surgical vantage point into an unfamiliar anatomical orientation. For this reason, practical training in the laboratory is necessary to adjust and expand the "comfort zone" of the surgeon.

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5 Juxtasellar Cisterns

Michael C. Huang

Keywords: epidermoid cyst, Kawase's quadrilateral, zygoma, interpeduncular cistern, chiasmatic cistern, petrosectomy

■ Case Presentation

A 42-year-old homeless man presented to the emergency department with complaints of headaches, nausea, and vomiting for 2 weeks. In the emergency department, he had a sudden decline in his mental status and suffered a witnessed tonic–clonic seizure. The patient was intubated for airway protection, and his seizure was arrested with boluses of Ativan. On follow-up examination, he was noted to have a left-sided hemiparesis. A computed tomography (CT) and magnetic resonance imaging (MRI) scan of the head were obtained.

Questions

1. What anatomical region does this mass occupy? Where do you think it originated?
2. What are the major differential diagnoses to consider?
3. What is the most likely growth pattern and the natural history of this lesion?

■ Diagnosis and Assessment

The CT showed a large hypodense extra-axial lesion measuring approximately 3 × 6 × 5 cm. The lesion appeared to be centered in the chiasmatic cistern and extended into the interpeduncular cistern and into the choroidal fissure. There was associated mass effect on the right temporal lobe, brainstem, the right lateral and third ventricles with up to 1 cm of midline shift. On MRI, the lesion appeared lobulated and was nonenhancing, hypointense on T1 (**Fig. 5.1a**), hyperintense on T2 (**Fig. 5.1b–d**), and hyperintense on diffusion-weighted imaging (DWI) (**Fig. 5.1e**). The lesion encased multiple intracranial arteries including the basilar artery, the proximal portions of bilateral posterior cerebral arteries, bilateral superior cerebellar arteries, and the right supraclinoid internal carotid artery (ICA).

The most likely diagnosis of this lobulated lesion that expanded the chiasmatic and interpeduncular cisterns and encased vessels is an epidermoid cyst. Intracranial epidermoid cysts are congenital inclusion cysts that comprise 0.2 to 1.8% of primary intracranial tumors. They are located off the midline and are most commonly found in the cerebellopontine angle cistern, followed by the fourth ventricle and the parasellar regions. On CT scan, epidermoid cysts appear as well-defined masses with similar attenuation as cerebrospinal fluid (CSF), and they do not enhance. On MR imaging, epidermoid cysts are isointense or slightly hyperintense to CSF signal on both T1- and T2-weighted images. They have incomplete suppression on fluid-attenuated inversion recovery (FLAIR) sequence. On diffusion-weighted sequences, epidermoid cysts show restriction and appear bright. Typically, there is no enhancement with contrast administration, though minimal rim enhancement occurs in approximately 25% of cases.

The major differentials for an epidermoid cyst include arachnoid cyst, dermoid cyst, neurocysticercosis, and cystic neoplasms. Arachnoid cysts will have similar signal characteristics as CSF on CT and on all MR sequences, including diffusion-weighted images. However, they typically displace rather than encase neurovascular structures. Dermoid cysts are four to nine times less common as epidermoid cysts. They typically appear in the midline and have imaging characteristics similar to fat rather than to CSF. Neurocysticercosis and cystic neoplasms will enhance with contrast administration and usually are associated with edema in the adjacent brain parenchyma.

Epidermoid cysts develop from ectodermal inclusion during neural tube closure between the third and fifth weeks of gestation. The glistening white cyst wall consists of stratified squamous epithelium, and the cysts enlarge with the desquamation of epithelium lining and accumulation of keratin and cholesterol breakdown products. Therefore, they grow slowly with a linear growth rate that is similar to the turnover of normal human skin. These tumors have a soft consistency that allows them to extend through the natural cleavage planes, filling and expanding sulci, fissures, and cisterns. They can extend through the incisura to occupy multiple cranial compartments. Epidermoid cysts surround and encase neurovascular structures, and clinical symptoms develop with compression of these critical structures, such as trigeminal neuralgia, diplopia, facial palsy, and hearing loss. Because of the slow growth rate of epidermoid cysts, symptoms associated with these tumors usually present only after the tumors have achieved a significantly large size. Clinical signs and symptoms may have prolonged courses prior to diagnosis.

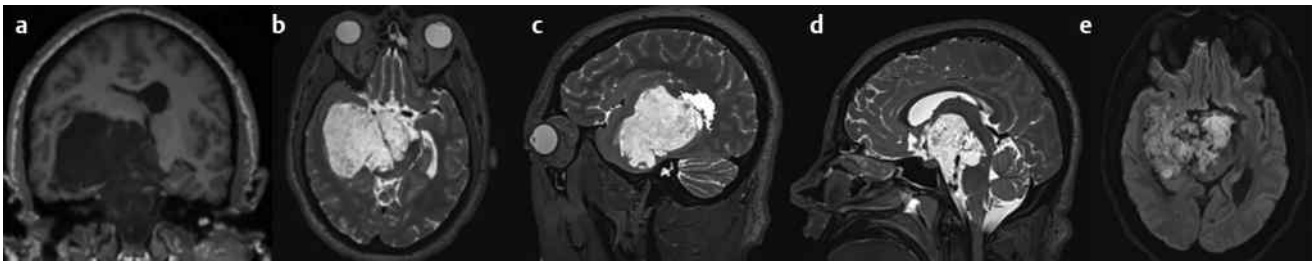


Fig. 5.1 Preoperative images. (a) T1 coronal, **(b)** T2 axial, **(c, d)** T2 sagittal, and **(e)** DWI images showing a large mass occupying the chiasmatic and interpeduncular cisterns, extending into the choroidal fissure. The signal characteristics coupled with the relationship with arteries make the most likely diagnosis an epidermoid.

■ Anatomical and Therapeutic Considerations

Since epidermoid cysts are benign tumors, the goal of surgery in most patients is total resection in order to avoid future recurrence. However, as the tumor spreads through the subarachnoid spaces, it can encompass and adhere to multiple neurovascular structures, such as the brainstem, cranial nerves, and perforating arteries. Leakage of cyst contents, which incites chemical meningitis, may contribute to the glue-like adhesive process. Aggressive dissection of tumor capsule from delicate neurovascular structures can result in significant morbidities and should be avoided. In such cases, surgical intervention should focus on alleviation of symptoms through conservative resection, with removal of cyst content and unattached portions of the cyst wall. As epidermoid cysts are congenital lesions and most patients become symptomatic during their fourth decade, recurrence from a subtotal resection may not require additional intervention for many years or decades.

The epidermoid cyst in this case occupied both supra- and infratentorial spaces (**Video 5.1**). In the cranial–caudal dimension, the lesion extended from the thalamus, hypothalamus, and the floor of the third ventricle, through the chiasmatic cistern and traveled anteriorly to the brainstem along the clivus through the interpeduncular cistern to just below the level of the internal auditory meatus. Anteriorly, the tumor displaced the optic chiasm superiorly but did not invade into the central skull base, and the pituitary gland and stalk were spared. Posteriorly, the tumor compressed the anterior margin of the midbrain and pons. The tumor extended laterally into the right choroidal fissure and invaded the temporal horn of the right lateral ventricle.

Options for Approach

As with any skull base operation, the surgical approach to epidermoid cyst resection must be designed to provide optimal visualization and surgical freedom while minimizing trauma to neurovascular structures. Epidermoid cyst contents are soft and readily amendable to aspiration. As internal debulking of the tumor progresses, an artificial “surgical channel” is created, providing an avenue to all the internal recesses of the tumor. The greatest risk for morbidity, however, resides in the dissection of the tumor capsule, which is often adherent to delicate neurovascular structures. Therefore, the approach must provide adequate exposure of the interphase between the capsule and vital structures. The approaches suitable for a transtentorial epidermoid cysts include subtemporal, retromastoid suboccipital, and combined supra- and infratentorial approaches performed in one or two stages.

In this case example, the approach must provide access to a large area, from the floor of the third ventricle cranially, to the internal auditory meatus caudally, to the anterior margin of the pons medially, and to the right choroidal fissure laterally. Several skull base techniques were combined to construct the approach. A frontotemporal orbitozygomatic (FTOZ) craniotomy would form the basic platform of the approach. This provides an anterolateral as well as a lateral subtemporal corridor to the interpeduncular cistern. The removal of the orbital rim flattens the trajectory along the anterior skull base and increases the degree of surgical freedom when working superiorly toward the third ventricle. The removal of the zygomatic arch allows the temporalis muscle to be retracted further inferiorly. This allows for more aggressive resection of the squamosal temporal bone to flatten the trajectory along the middle fossa floor (**Fig. 5.2**).

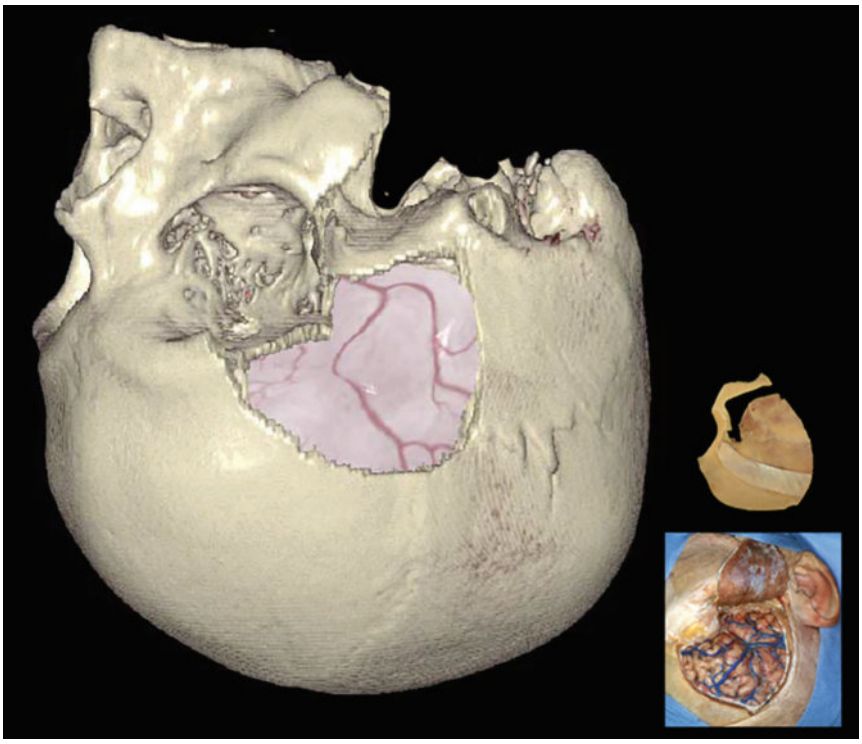


Fig. 5.2 Illustration of right-sided orbitozygomatic craniotomy opening. The wide opening accesses both the anterolateral and lateral corridors. Removal of the orbital rim and zygomatic arch increases the surgeon’s ability to look in an inferior-to-superior trajectory. In addition, with the arch detached, the temporalis can be retracted inferiorly to flatten the line of sight along the middle fossa floor. Inset: the bone flap of the orbitozygomatic craniotomy and the opening demonstrated in a cadaveric dissection with the dura removed.

Supratentorial Considerations

The FTOZ craniotomy can be performed using either a one- or two-piece technique. In the two-piece technique, a standard pterional craniotomy is fashioned first and the orbitozygomatic osteotomy is performed separately. In the one-piece technique, the creation of a MacCarty keyhole and the inferior orbital fissure are used as communicating points for the orbital osteotomy cuts in order to complete the entire craniotomy as a single bone flap. Conceptually and technically, the two-piece technique is simpler. It potentially allows for the preservation of larger portions of the orbital roof and lateral orbital wall in order to minimize postoperative enophthalmos. The attachment of the masseter muscle to the zygomatic arch can be preserved when using a two-piece technique in order to avoid postoperative issues with mastication. However, compared with the one-piece approach, the two-piece technique necessitates an osteotomy across the frontal bone on the forehead, that is, the separation line between the two pieces, with potentially undesirable cosmetic outcomes. The bone reconstruction of two pieces of bone flaps at the end of the operation may also damage cosmesis. Ultimately, the decision between a one- and two-piece technique is dependent on surgeon proficiency and preference. Both techniques can be mastered and performed with excellent results and minimal complications.

Infratentorial Considerations

Finally, the surgical approach for this epidermoid must reach into the posterior fossa for the infratentorial portion of the tumor. While a subtemporal corridor can provide a path to the posterior fossa by division of the tentorium, the temporal lobe must be retracted away from the middle fossa floor to "create" space in the corridor. This puts bridging temporal veins at risk, which may result in venous infractions, especially if the vein of Labbé is injured. Furthermore, even with the tentorium divided, the working angle through a subtemporal approach remains limited by the petrous bone edge laterally.

Approach of Choice

To avoid these potential problems, an anterior petrosectomy would be added to the FTOZ. By resecting the anterior petrous apex, the posterior fossa would be accessed through the Kawase's quadrilateral, which is bounded anteriorly by the mandibular division of the trigeminal nerve, posteriorly by the arcuate eminence, laterally by the greater superficial petrosal nerve (GSPN), and medially by the superior petrosal sinus along the petrous edge (**Fig. 5.3**). Containing no vital structures, this block of bone would be removed to reach the upper portion of the clivus from the dorsum sellae to the level of the internal auditory canal (**Fig. 5.4**). In addition to widening the operative window, the anterior petrosectomy also shortens the working distance to midline structures and mitigates the retraction time and forces on the temporal lobe.

For the reasons cited above, the plan was made for a one-piece FTOZ craniotomy, with an anterior petrosectomy, for a maximally safe removal of the tumor.

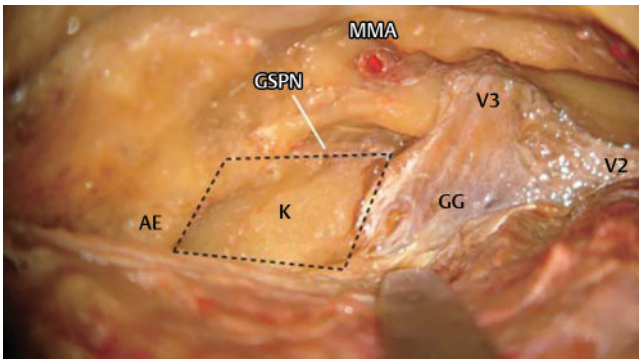


Fig. 5.3 Cadaveric dissection of the left middle fossa floor. Kawase's quadrilateral (K) is bordered by V3, the greater superficial petrosal nerve (GSPN), arcuate eminence (AE), and the petrous ridge. GG, gasserian ganglion; MMA, middle meningeal artery. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

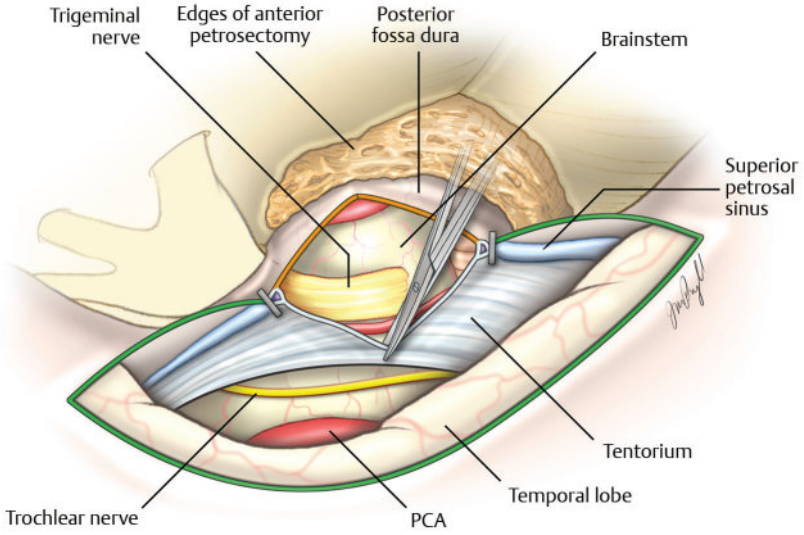


Fig. 5.4 View of the right posterior fossa through the window of the anterior petrosectomy. The subtemporal dura was opened parallel to the floor of the middle fossa. The green edges denote this cut. The posterior fossa dura was opened perpendicular to it, and the orange edges denote this cut. These two incisions joined at the superficial petrosal sinus. The sinus was ligated and cut, which started the incision of the tentorium at its lateral edge. This cut was carried medially until the tentorium was cut completely to the incisura.

Questions

1. What cranial nerve functions are at risk during an anterior petrosectomy?
2. What is the function of the inferior orbital fissure during a one-piece FTOZ craniotomy?

■ Description of the Technique

Soft Tissue Dissection

At the beginning of surgery, a left frontal external ventricular drain (EVD) was placed at Kocher's point to assist with brain relaxation during the extradural dissection for the anterior petrosectomy. The patient's head was rotated approximately 30 degrees to the left. Baseline neurophysiological monitoring was established, including monitoring for trigeminal, facial nerve and for brain auditory-evoked responses (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: anterolateral

Craniotomy: orbitozygomatic

Modifier: zygomatic osteotomy, anterior petrosectomy

A single curvilinear incision was fashioned from the widow's peak and extended posteriorly to end just anterior to the tragus. The scalp was elevated leaving the pericranium attached to bone. The superficial layer of the deep temporal fascia was incised from the root of the zygoma to the pterion. Dissection was carried through the fat pad down to the deep temporal fascia, and an interfascial dissection was performed to expose the entire zygomatic arch, while protecting the frontal branch of the facial nerve (**Fig. 5.5a**). Dissection was carried anteromedially to expose the entire zygomatic bone and the lateral aspects of the orbit. The superior orbital rim was exposed medially to the supraorbital notch, and the periorbita was dissected free from the inner surface of the orbit from the supraorbital notch down to the inferior orbital fissure. The temporalis muscle was cut along the superior temporal line while preserving a 1 cm myofascial cuff superiorly to be used for closure. Perpendicular to this, it was also cut along the skin incision and then reflected inferiorly, exposing the pterion and the inferior orbital fissure (**Fig. 5.5b**).

One-Piece Frontotemporal Orbitozygomatic Craniotomy

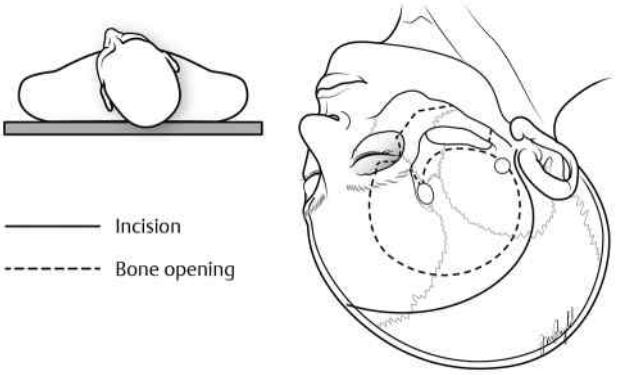
Operative Setup

Position: supine, head turned 30 degrees

Incision: curvilinear, tragus to across midline

Craniotomy: one-piece frontotemporal orbitozygomatic craniotomy

Durotomy: C-shaped based on pterion



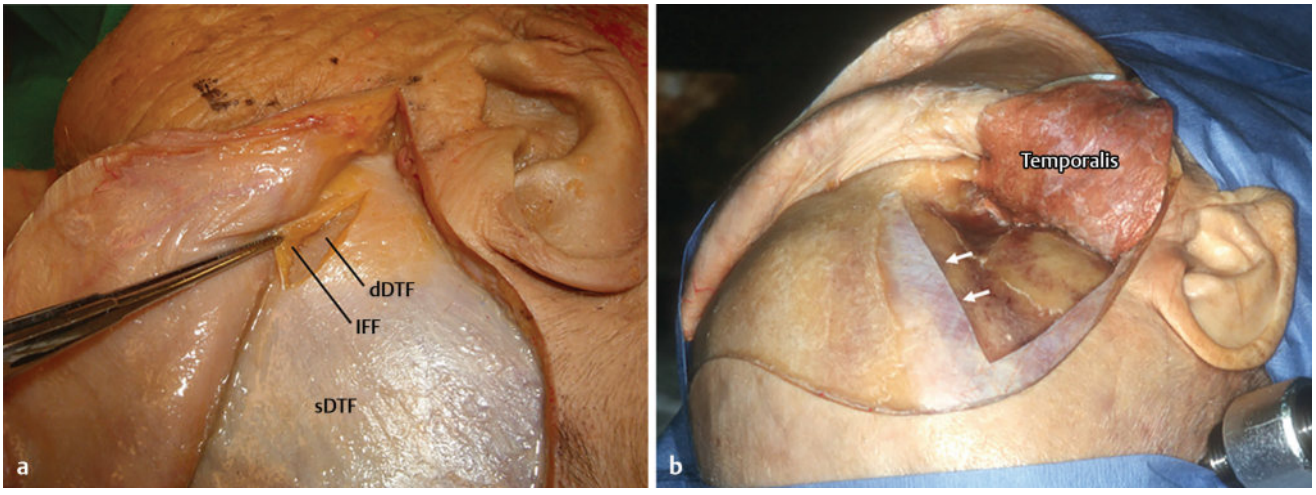


Fig. 5.5 Cadaveric dissection showing the handling of the soft tissue before the frontotemporal orbitozygomatic craniotomy. (a) Incision is made in the superficial layer of deep temporal fascia to expose the deep temporal fat. The superficial layer of the temporal fascia is separated from the deep layer and the superficial layer is raised along with the scalp to protect the frontal branch of the facial nerve. dDTF, deep layer of the deep temporal fascia; IFF, interfascial fat pad; sDTF, superficial layer of the deep temporal fascia. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.) **(b)** The temporalis muscle is reflected inferiorly after a right-angled incision is made. *White arrows*, myofascial cuff approximately 1 cm thick just below the superior temporal line. This cuff will be used for reapproximation of the muscle during closure.

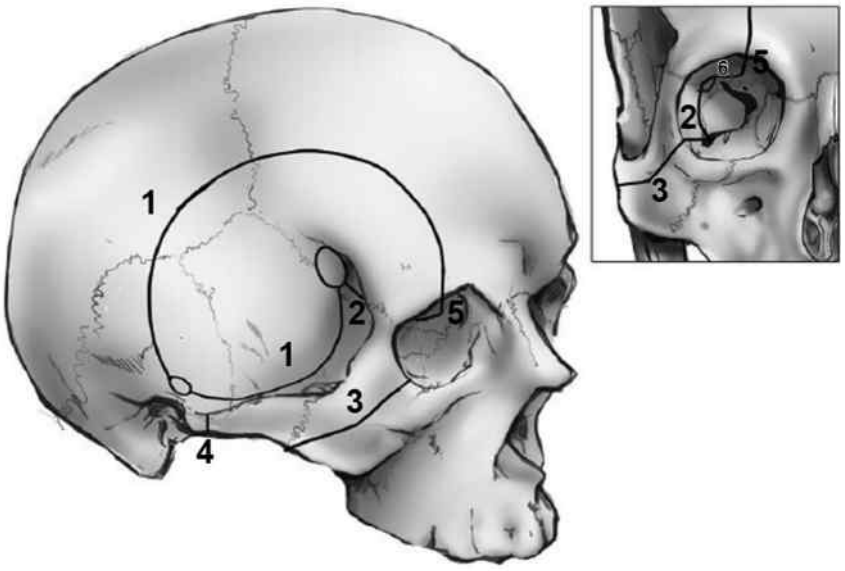


Fig. 5.6 The six cuts required to perform the one-piece frontotemporal orbitozygomatic craniotomy. (Reproduced from Di Ieva A, Lee JM, Cusimano MD. Handbook of Skull Base Surgery. 1st ed. Thieme; 2016.)

At this point, attention was turned toward performing the one-piece FTOZ. First, a MacCarty keyhole burr hole was placed over the frontosphenoidal suture approximately 1 cm posterior to the frontozygomatic junction. The frontal dura was exposed in the superior portion of the burr hole, the periorbita in the inferior portion, while the orbital roof separated the two halves. A temporal burr hole was placed adjacent to the root of the zygoma (see **Operative Setup**).

The one-piece FTOZ was fashioned using six bone cuts (**Fig. 5.6**).

1. With a craniotome and footplate, a bone cut was made in the frontal portion of the MacCarty burr hole, posteriorly across the sphenoid ridge, to reach the temporal burr hole. From there, the cut extended superiorly and then anteriorly to the supraorbital ridge, ending just lateral to the supraorbital notch.
2. Using the orbital portion of the MacCarty burr hole, the craniotome/footplate cut inferiorly along the lateral orbital wall to exit the anterolateral part of the inferior orbital fissure.
3. Removing the footplate, the drill cut across the body of the zygoma just above the zygomaticofacial foramen and down to the anterolateral edge of the inferior orbital fissure.
4. A cut was made across the posterior end of the zygomatic root, thereby completely disconnecting the zygoma.
5. While protecting the periorbita, the supraorbital rim was cut lateral to the supraorbital notch. This cut was then extended back along the orbital roof with a small osteotome for at least 1 cm.
6. The sixth and final cut was made using the osteotome to cut across the orbital roof from the MacCarty burr hole to connect with the medial orbital bone cut.

Once the bone flap was elevated, additional bone from the anterolateral portion of the orbital roof was removed and the lesser sphenoid wing was resected down to the meningo-orbital band. The frontal sinus was not violated.

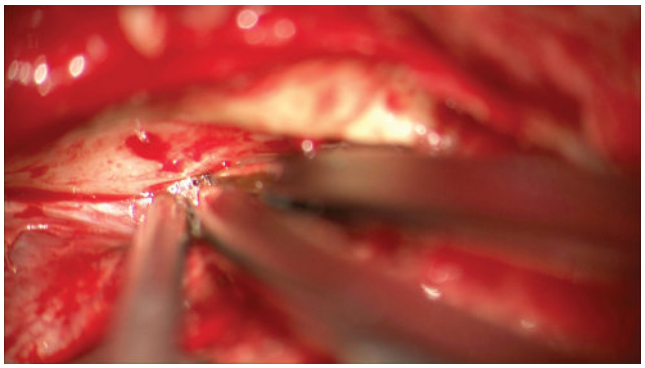


Fig. 5.7 Intraoperative image. The middle meningeal artery is coagulated and then cut at the foramen spinosum. This release maneuver allowed the mobilization of the subtemporal dura off the floor of the middle fossa all the way to the petrous apex.

Anterior Petrosectomy

The patient was rotated further to the left to bring the superior sagittal sinus more parallel to the floor. With microscope visualization and CSF drainage from the EVD for brain relaxation, the middle fossa temporal lobe dura was elevated in a posterior-to-anterior direction to avoid stretching the GSPN. The middle meningeal artery was divided as it exited the foramen spinosum in order to carry the dissection to the petrous edge (**Fig. 5.7**). Landmarks identified on the middle fossa floor included the mandibular division of trigeminal nerve passing through foramen ovale anteriorly, the GSPN, and the arcuate eminence posteriorly. The GSPN nerve was stimulated with the facial nerve stimulator for confirmation (**Fig. 5.8**).

The angle between the arcuate eminence and the GSPN was bisected to approximate the position of the internal auditory canal. The roof of the arcuate eminence was drilled off and the superior canal could be faintly seen through the residual bone (i.e., blue-lined). With that landmark in mind, the petrous

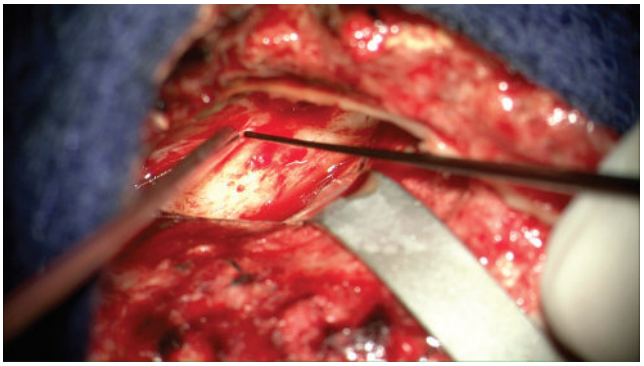


Fig. 5.8 Intraoperative image. With the facial nerve stimulator probe, the position of the greater superficial petrosal nerve (GSPN) was confirmed.

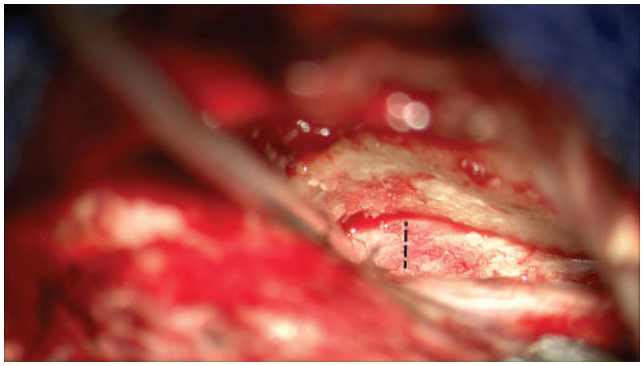


Fig. 5.9 Intraoperative image. As seen through the area of the anterior petrosectomy, the posterior fossa dura is cut (*dashed line*). The subtemporal dura was simultaneously incised parallel to the floor of the middle fossa. The point where these two perpendicular cuts joined was the superior petrosal sinus. This was coagulated and cut as well, thus starting the incision of the tentorium laterally.

ridge was drilled down until the porus of the internal auditory canal was seen. Once the internal auditory canal porus was visualized, the petrous ridge was taken down anteriorly to the Meckel cave. This resulted in exposure of the dura of the posterior fossa. More laterally, the bone was resected up to the GSPN. Posterolaterally, bone over the cochlea was left intact which was critical in preserving hearing.

Tumor Resection

The dura was opened in a C-shaped fashion with the pedicle based on the sphenoid ridge. The most superficial part of the tumor was identified, which was over the middle temporal gyrus. A small cortisectomy was performed in the middle temporal gyrus and the lateral portion of the tumor was encountered. The tumor capsule was white and glistened with a sheen similar to that of mother-of-pearl. The inside of the tumor contained waxy and flaky material that was soft and was easily aspirated. Resection of the tumor carried the dissection toward midline and the apex of the basilar artery was encountered. Tumor was meticulously dissected away from both posterior cerebral arteries and superior cerebellar arteries, as well as from all associated perforators. The right posterior communicating artery was traced retrograde from its insertion on the right posterior cerebral artery to the right ICA. Great care was taken to preserve all arteries and perforators. Bilateral oculomotor nerves were identified and protected.

As the tumor resection continued inferiorly, the surgical corridor became increasingly narrower and more restrictive. Therefore, the direction of approach was shifted in order to take advantage of the anterior petrosectomy. The patient was rotated to the left once more and the anterior petrosectomy site was identified. Perpendicular dural cuts were made in the middle fossa and posterior fossa dura with the superior petrosal sinus in between (**Fig. 5.9**). The superior petrosal sinus was coagulated and cut. The tentorium was cut toward midline, making sure to avoid injury to the trochlear nerve. The cut tentorial edges were retracted and pearly white tumor could be visualized in the interpeduncular cistern (**Fig. 5.10**). The soft tumor was aspirated until the basilar artery was visualized. The resection of the tumor then extended inferiorly until facial and vestibulocochlear nerves were identified. Facial nerve activity was confirmed with direct stimulation. Additional tumor would be visualized inferior to the VII/VIII nerve complex but was left in situ in order to avoid injuries to the facial nerve

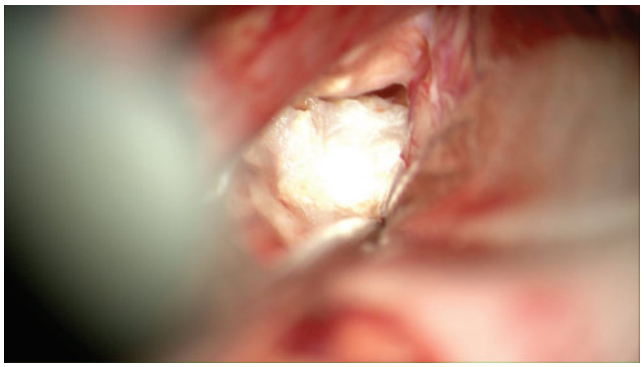


Fig. 5.10 Intraoperative image. The epidermoid tumor was seen in the interpeduncular cistern through the dural incisions described in **Fig. 5.9**.

At this point, attention was turned toward the superior portion of the tumor. Working once more through the temporal corridor that had been developed from tumor debulking, dissection was turned toward the hypothalamus and the floor of the third ventricle. The cyst contents were easily aspirated. However, small remnants of tumor capsule were tightly adherent to the hypothalamus. These were left in place in order to avoid injury to underlying neural tissue. The dissection continued across the floor of the third ventricle toward the contralateral lateral ventricle. At this point, with the exception of small pieces attached to the hypothalamus, all visible tumors superiorly, laterally, and medially had been removed. Inferiorly, there was tumor below the facial nerve and anterior to the medulla, which was not assessable from the current supratentorial approach. The decision was made to proceed with closure.

Closure

The entire surgical field was irrigated with copious amounts of irrigation. Additional flakes of tumor floated free with the irrigation fluid and were removed. The frontotemporal dura was closed in a watertight fashion. The temporal and posterior fossa

dura adjacent to the anterior petrosectomy were reapproximated. A small piece of temporalis muscle was harvested and placed over the durotomy site. TISSEEL was applied to reinforce the reconstruction. The one-piece FTOZ flap was secured using titanium plates and screws. A single subgaleal drain was placed and the wound was closed in layers.

Surgical Pearls

1. A thorough understanding of the MacCarty burr hole and the inferior orbital fissure is critical for mastery of the one-piece FTOZ approach.
2. Resection of the petrous apex in an anterior petrosectomy approach provides exposure from the dorsum sellae to the internal auditory canal.
3. Brain relaxation is critical during extradural dissection and bone resection for anterior petrosectomy. A lumbar drain, or an EVD may be necessary.
4. The bone covering the ICA on the floor of the middle fossa may be thin or absent. Care must be taken during elevation of the temporal dura and during anterior petrosectomy to avoid injuring the petrous segment of the ICA.
5. When dividing the tentorium, be sure to avoid the trochlear nerve, which travels along the edge of the tentorium at the incisura.

■ Aftercare

The patient was admitted to the intensive care unit for postoperative care. He was kept intubated given the longer duration of the operation. His EVD was kept open to drain. Steroids were administered in the postoperative period to reduce the risk for aseptic meningitis due to the spillage of cyst content during tumor resection. Given that remnants of tumor capsule were purposely left behind to avoid hypothalamic injury and residual tumor was seen anterior to the brainstem on postoperative imaging (**Fig. 5.11**), serial radiographic surveillance will be required. Malignant transformations of epidermoid cysts have been reported, and is postulated to be associated with chronic inflammation secondary to irritation from leakage of cyst content, such as during surgical resection.

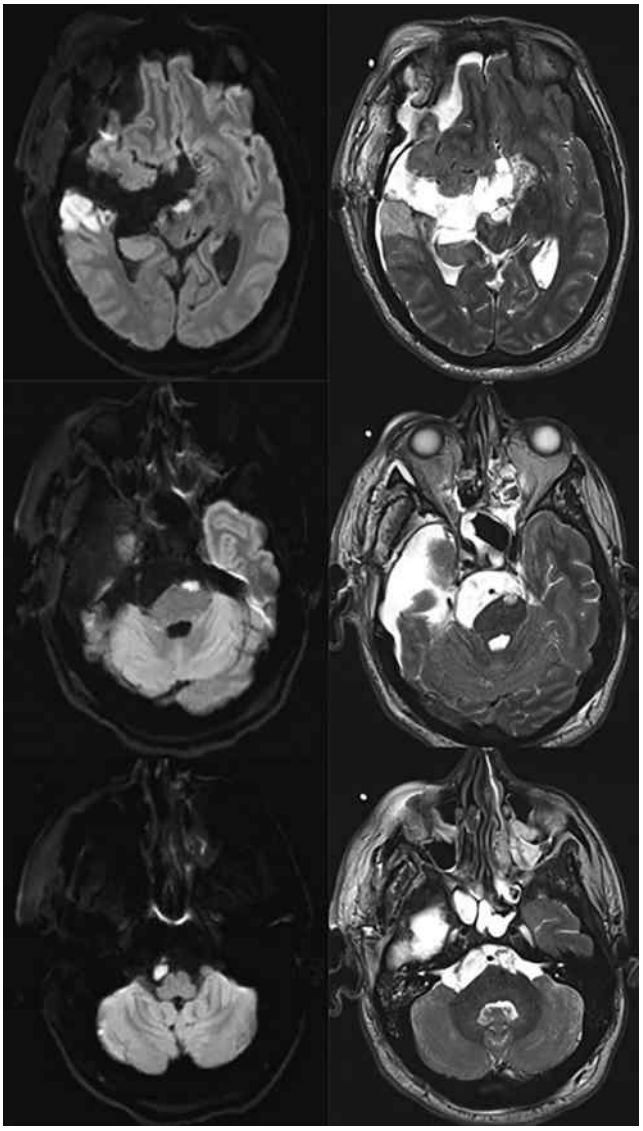


Fig. 5.11 Postoperative MRI. DWI (*left column*) and T2 (*right column*) showed a small remnant anterior to the brainstem.

Perspective

Wenya Linda Bi and Ossama Al-Mefty

■ Introduction

Epidermoid cysts conform to an epitaph that can be prescribed for skull base tumors in general: *with recurrence, the price is high*. More so than for its skull base compatriots, for which adjuvant radiation and limited pharmacotherapies remain options, surgery is the only treatment for epidermoid cysts. Among de novo cases, true total removal of the epidermoid capsule leads to definitive cure, whereas presence of residual capsule nearly guarantees recurrence in the long term. In both first-time surgery and surgery for recurrence, resection improves the quality of life as measured by the Karnofsky Performance Scale. However, each subsequent recurrence increases the difficulty of the operation, the number of loculations observed, and the risk of inadvertent neurological injury, as the arachnoid planes between capsule and neurovascular structures become progressively obscured. Furthermore, a subtotal removal strategy is associated with higher rates of aseptic meningitis, hydrocephalus, and indeed, mortality, as compared to intended total removal.

Thus, every effort should be made for a total removal, especially in de novo cases. Critical for the achievement of the goal for total resection are meticulous microsurgical technique with bimanual dissection of the capsule from the arachnoid plane investing nerves and vessels, "skull base" approaches for optimal exposure, intraoperative neurophysiological monitoring, and endoscopic assistance in seeing and working around "blind" corners. The choice of surgical approach is dictated by the extension of the lesion, its perceived consistency and adherence, existing preoperative deficits, and the venous anatomy encountered. When necessary, multiple skull base approaches may be indicated to afford an optimal, multidirectional access.

Despite such effort, and the awareness of inevitable recurrence with subtotal resection, capsular adherence to critical neurovascular structures—including perforator arteries, cranial nerves, and the brainstem—can at times stymie the dedicated pursuit of total resection. The decision of how to operate on a recurrent epidermoid becomes much more nuanced, as the acceptable risk/balance ratio diverges between individual patients.

■ Case Presentation

A 35-year-old man presented with 6 months of dizziness, vertigo, difficulty swallowing, and deteriorating gait. Evaluation was notable for aspiration on video swallow testing and moderate right-sided hearing loss. His extraocular movements remained intact, as did his facial strength, and power in his extremities. MRI of the brain revealed a 6-cm T1-hypointense, T2-hyperintense mass with restricted diffusion, extending from the suprasellar region into the posterior fossa level, with severe brainstem compression (**Fig. 5.12**).

■ Anatomical and Therapeutic Considerations

The MRI characteristics of the lesion are consistent with an epidermoid cyst, which frequently has the insidious ability to grow to remarkable extent before presentation, crossing arachnoid trabeculations and multiple skull base compartments to involve both the supra- and infratentorial space. This case illustrates one such giant epidermoid, which extended from well above the optic chiasm in the chiasmatic cistern into the posterior fossa. The mass involved the right cerebellopontine region and extended to the level of the jugular foramen. Near the midline, it crossed the interpeduncular cistern to the contralateral ventral brainstem.

Approach of Choice

The radiographic evidence of multiple cranial nerve involvement, and the degree of ventral extension to the contralateral side, dictated the need for posterolateral exposure into the posterior fossa. However, the challenge of reaching from this exposure to the supratentorial space, without jeopardizing critical venous drainage in the tentorium, prompted a separate middle fossa exposure, which can be achieved through a preauricular zygomatic approach. Thus, a posterior petrosal approach (retrolabyrinthine posterior petrosectomy), combined with preauricular, zygomatic middle fossa craniotomy for an anterior petrosal approach (anterior petrosectomy/Kawase), was therefore chosen to allow maximal access to the tumor from its rostral-to-caudal extent.

A combined petrosal approach traditionally implies sectioning of the tentorium and the superior petrosal vein. Not infrequently, however, critical veins draining the inferior temporal lobe may insert into the superior petrosal vein, and interruption of this drainage may have devastating consequences, especially in the dominant temporal lobe. Therefore, a preoperative dynamic CT angiography or CT venography was obtained to visualize the venous anatomy in the area of the tumor and the intended approach. Low-lying inferior temporal veins and the insertion of the vein of Labbé must be carefully noted and these veins must be preserved during the exposure. For soft, suckable lesions such as an epidermoid, the labyrinth and middle ear apparatus are generally preserved for patients with intact hearing. The assistance of the endoscope to reach the ventral and contralateral extension further relegates the translabyrinthine or transcochlear approach from consideration.

Despite the presence of overt hydrocephalus with transependymal flow, preoperative CSF diversion was deferred, as

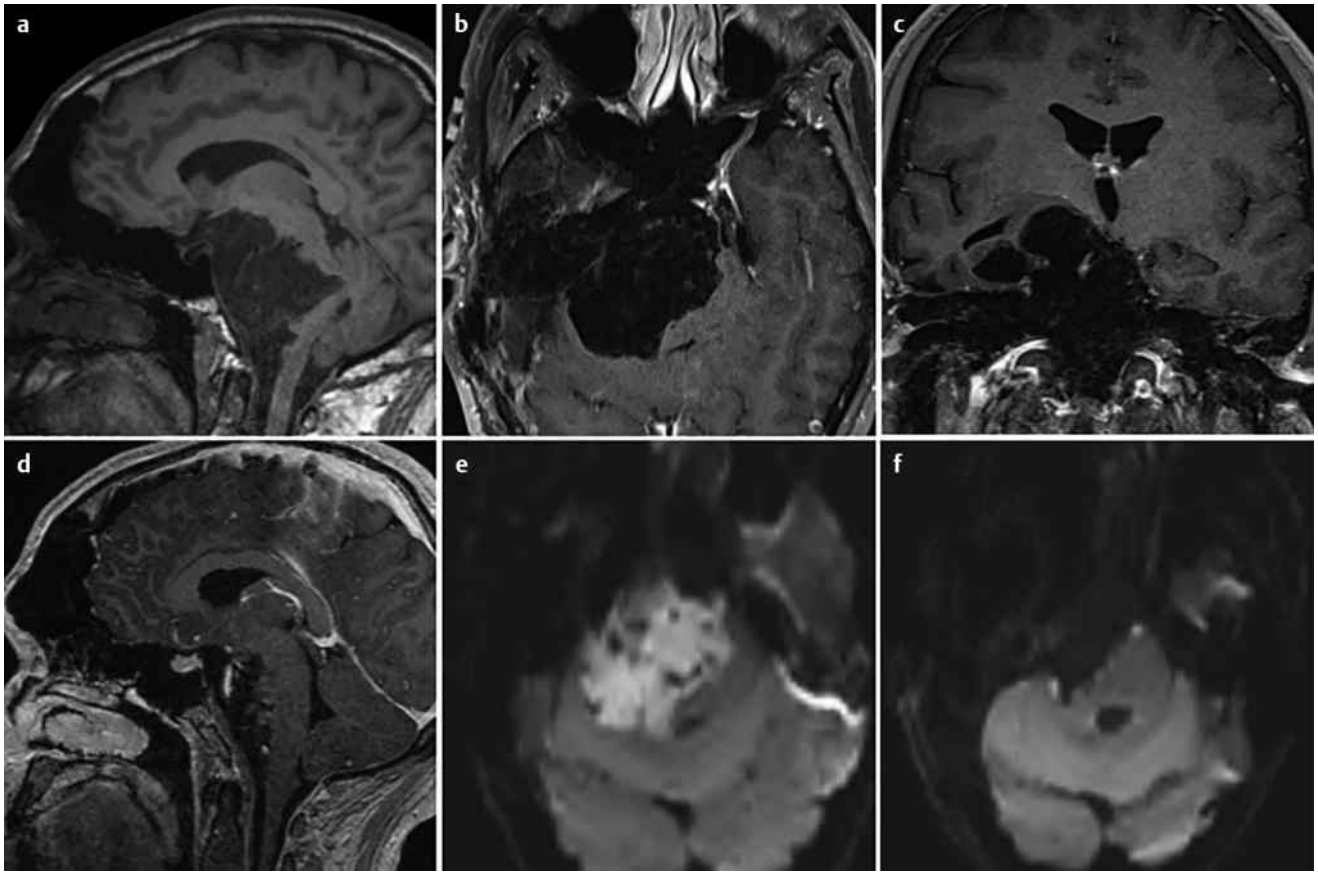


Fig. 5.12 Preoperative T1-weighted contrast-enhanced MRI in the **(a)** sagittal, **(b)** axial, and **(c)** coronal planes demonstrate a large hypointense mass extending from the suprasellar region to the level of the jugular foramen, causing severe compression of the brainstem. **(d)** Postoperative sagittal T1-weighted contrast-enhanced MRI demonstrating complete resection of the epidermoid cyst. **(e)** Preoperative and **(f)** postoperative axial diffusion-weighted MRI demonstrating restricted diffusion throughout the epidermoid, with subsequent resolution after surgery.

the thought was that with definitive removal of the tumor, the obstructive hydrocephalus would resolve with the elimination of its cause. The concept of minimizing postoperative complications by leaving tumor capsule adherent to critical structures may prove counterproductive, as it ensures eventual recurrence, the treatment of which would have higher surgical morbidity. Foreseeing this future risk, diligent upfront pursuit of capsular removal is the prudent choice. Careful preservation of the arachnoid plane between the cyst wall on one side and neurovascular structures on the other provides the means to attempt, and hopefully achieve, total resection, which was our goal.

■ Description of the Technique

The patient was positioned supine, with the ipsilateral shoulder bolstered by a roll. His head was rotated 15 degrees to the opposite side, chin slightly flexed, and vertex tilted to the contralateral shoulder, to allow visualization above and below the petrous ridge. Electrodes were placed for neuromonitoring of cranial nerves III, V, VI, VII, X, XI, and XII, and somatosensory-evoked potentials. The scalp was incised in the fashion of a shepherd's hook, starting from 1 cm anterior to the tragus, traveling superiorly and curving backward in the temporal area above the ear, and then in curvilinear fashion behind the mastoid in the postauricular space, maintaining a wide base to preserve vascular supply to the skin flap, which is retracted inferiorly and anteriorly. The temporalis fascia was sharply incised and separated from the underlying temporalis muscle and reflected caudally in continuity with the sternomastoid muscle to maintain a vascularized flap for reconstruction purposes. The zygoma was freed from fascial attachments along its superior surface and sectioned at its most anterior and posterior points. The temporalis muscle was then dissected in subperiosteal fashion and reflected inferiorly and anteriorly to expose the base of the middle fossa. Four pairs of burr holes were placed, two pairs abutting the transverse sinus, one anteriorly along the base and frontal limits of the temporal bone, and one inferiorly in the posterior fossa. Following epidural dissection, craniotomy cuts were made in the supra- and infratentorial compartments, while the burr holes spanning the transverse sinus were connected from the surface of the bone to the level of the sinus using a cutting drill bit (**Fig. 5.13**).

The mastoid cortex was harvested for eventual reconstruction and an extensive mastoidectomy and posterior petrosectomy was performed to skeletonize the sigmoid sinus and remove the posterior petrous ridge, while preserving the labyrinth and facial canal (**Fig. 5.14**). A small dural incision is made in the presigmoid dura to release CSF and attention was turned to the middle fossa. Dissection was performed extradurally along the middle fossa floor until the GSPN, arcuate

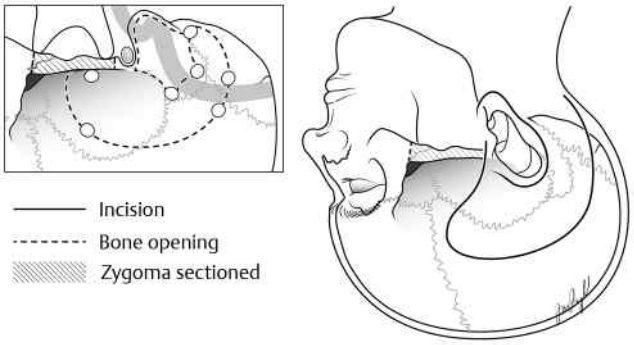


Fig. 5.13 Incision and bone opening. The scalp was incised in the fashion of a shepherd's hook, starting from 1 cm anterior to the tragus, traveling superiorly and curving backward in the temporal area above the ear, and then in curvilinear fashion behind the mastoid in the postauricular space. The zygoma was freed from fascial attachments along its superior surface and sectioned at its most anterior and posterior points. Two pairs of burr holes were placed abutting the transverse sinus, one anteriorly along the base and frontal limits of the temporal bone, and one inferiorly in the posterior fossa. The craniotomy cuts were made in the supratentorial and infratentorial compartments, while the burr holes spanning the transverse sinus were connected from the surface of the bone to the level of the sinus using a cutting drill bit.

eminence, lateral wall of the cavernous sinus, and mandibular nerve sheath were visualized, with the petrous apex (Kawase's quadrilateral) bounded therein. The middle meningeal artery was coagulated and spinosum plugged with bone wax. The petrous apex was drilled with a diamond drill bit (anterior petrosectomy) until posterior fossa dura could be visualized. Dura was further opened in the presigmoid posterior fossa and along the base of the temporal lobe, connecting at the distal superior petrosal sinus and lateral tentorium, away from the insertion of any critical temporal veins.

Full exposure of the tumor was afforded by this combined petrosal approach, revealing a pearly shellac-like substance, which was removed in stepwise fashion (**Fig. 5.15**). As important as the skull base exposure is the microscopic handling of the epidermoid capsule. Bimanual dissection under high magnification offered the best chance for extricating the cyst capsule from adhered cranial nerves, perforator vessels, brainstem, and other neural structures. As such, cranial nerves VII, VIII, followed by the lower cranial nerves were individually inspected, with tumor removed from their investing arachnoid. Dissection of the tumor medially exposed the basilar artery and its perforators, which were meticulously preserved, as was the brainstem surface. The tumor was followed across the midline to the contralateral aspect of the brainstem across the ventral pons and midbrain. The abducens nerve, up to its insertion at Dorello's canal, and the trigeminal nerve were sequentially encountered and followed rostrally. The oculomotor and trochlear nerves, as well as the posterior cerebral artery and carotid artery, were encountered in the interpeduncular cistern. A substantial portion of the tumor resided in the suprasellar space, causing compressive hydrocephalus against the third ventricle and hypothalamus. This portion was removed last.

Given the propensity of epidermoids to infiltrate across skull base compartments, around cranial nerves, and into dural folds or osseous foramina, areas of suspected residual must

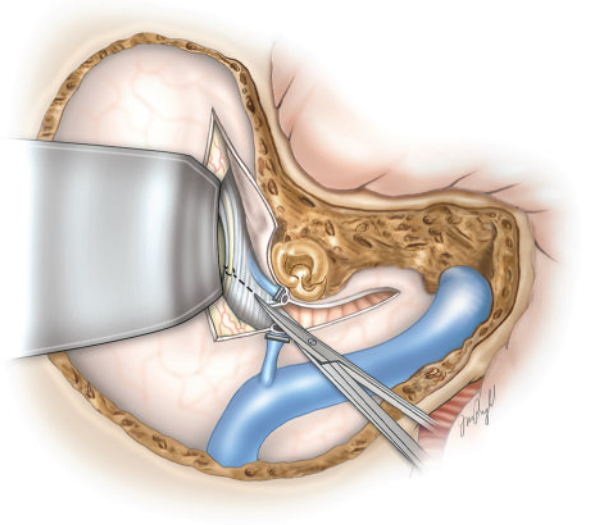


Fig. 5.14 Retrolabyrinthine posterior petrosectomy. The posterior petrosectomy was performed to skeletonize the sigmoid sinus and remove the posterior petrous ridge, while preserving the labyrinth and facial canal. The distal superior petrosal sinus was ligated and cut. The incision of the tentorium was made parallel to the superior petrosal sinus toward the incisura, ending posterior to the entrance of the trochlear nerve into the tentorium. Together with the opening of the anterior petrosectomy (**Fig. 15.4**), the combined petrosal opening provided a wide exposure to both supratentorial and infratentorial components of the epidermoid cyst.

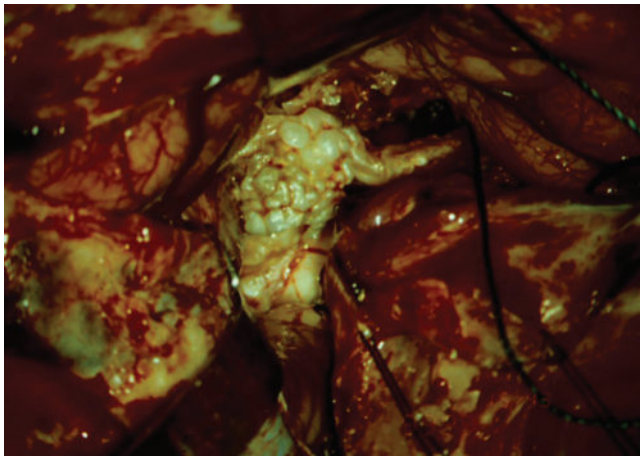


Fig. 5.15 Intraoperative view of giant epidermoid cyst with white pearly substance, with exposure offered by a combined anterior and posterior petrosal approach.

be systematically interrogated. The endoscope was therefore introduced to inspect blind spots during the removal of giant epidermoids, which included extension to the contralateral ventral brainstem, Meckel's cave, and ventral to the facial and lower cranial nerves.

After having achieved the goals of intended resection, abdominal fat graft is used to plug the petrous apex and aerated mastoid, bolstered by a piece of pericranial graft to protect the exposed trigeminal nerve at Meckel's cave. Dura was grafted with banked fascia lata. A tongue of the previously preserved

vascularized temporalis fascia graft was rotated in along the drilled petrous ridge and apex to further thwart CSF leak. The temporal dura was then tacked up to obliterate large epidural voids, and the calvarial vault was reconstructed with the harvested bone flap, mastoid cortex, and hydroxyapatite.

Despite meticulous surgical resection, spillover of epidermoid cyst contents may provoke a florid chemical meningitis. Intra- and postoperative steroids are therefore employed to reduce the incidence of such. Excessive irrigation was avoided during the resection to prevent distal spread of cyst contents throughout the subarachnoid space.

Postoperatively, the patient developed a transient abducens weakness. Formal swallow evaluation 1 week after surgery revealed no signs of aspiration and resolution of his dysphagia.

Commentary

Skull base approaches permit establishment of wide and flexible corridors, conducive to exploration of giant epidermoid cysts, which frequently invest along multiple compartments of the cranial base. Diligent pursuit of the tumor capsular removal along arachnoid planes affords the best opportunity to achieve total removal. The use of endoscopes, combined with microscopic magnification, allows visualization behind blind corners where epidermoids have a propensity to creep. Given the significantly higher morbidity and mortality associated with resection of recurrent epidermoid cysts, the goals of surgery for recurrent cases may need to be tailored depending on the clinical status, comorbidities, and age of the patient. Symptomatic decline may compel operating on a recurrent epidermoid cyst more so than radiographic progression alone. In elderly patients, decompression of cyst contents may allow for years of symptomatic relief. The opportunity for total removal of the capsule is best for de novo cases, while on recurrence, the chance for success is much less because of the adhesions and compartmentalization as a result of the first operation.

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6 Cavernous Sinus

Georgios A. Zenonos and Juan Carlos Fernandez-Miranda

Keywords: cavernous sinus, Kawase, Hakuba, Dolenc

■ Case Presentation

A 53-year-old otherwise healthy woman presented to a community hospital with sudden onset of numbness and tingling in the right forearm. She also endorsed a 2-month history of subtle diplopia with right gaze or up gaze. On physical examination, the patient was found to have a partial right abducens palsy, as well as a pupil-sparring oculomotor nerve palsy with some ptosis. There was some proptosis on the right. She had a history of right-sided amblyopia, and her visual acuity on examination was OD 20/30, OS 20/25. Magnetic resonance imaging (MRI) was obtained, which showed a homogeneously enhancing mass lesion involving the cavernous sinus on the right, as well as the tentorium, extending into the medial middle fossa floor, Meckel's cave, the crural cistern, the ambient cistern, and the lateral prepontine cistern in the posterior fossa. A computed tomography (CT) angiogram showed that the carotid artery was encircled and narrowed by the tumor (**Fig. 6.1**). The patient also had a CT of the chest, abdomen, and pelvis without any significant findings.

Questions

1. How does the differential diagnosis of the patient's lesion influence the next step in her management?
2. What anatomical structures and compartments are involved to explain the patient's symptoms?

■ Diagnosis and Assessment

The most likely diagnosis for this mass is a sphenocavernous–tentorial meningioma. The dural tail and the homogenous enhancement are virtually diagnostic, practically ruling out all other possibilities. For academic purposes, the differential diagnosis should include metastasis (which was even less likely since the whole-body CT scan was negative for any other malignancy), as well as inflammatory lesions, such as sarcoid, which is very rare.

With the leading diagnosis being meningioma, the management options discussed with the patient were the following:

1. Aggressive surgical resection with curative intent: This would require balloon test occlusion (BTO) to assess risks of internal carotid artery (ICA) sacrifice and potential need for revascularization, and would carry high risk of permanent cranial nerve palsies. In fact, tumor involvement of cranial nerves might require their surgical sacrifice for complete tumor resection.
2. Tailored surgical resection with the goal of maximizing tumor resection while minimizing surgical morbidity: The goal would be to remove all the tumor except the component within the cavernous sinus and Dorello's canal, including resection of the tentorial component, posterior fossa, middle fossa, with decompression of the cavernous sinus and optic canal. The residual tumor at the cavernous sinus can then be treated with any of the options 3 to 5.
3. Fractionated radiation therapy, as a sole treatment or after tailored resection.
4. Radiosurgery, as a sole treatment or after tailored resection.
5. Continued observation.

Given the relatively young age, the large tumor volume, and the presence of cranial neuropathies causing constant diplopia, we recommended surgical resection of the lesion. Continued observation of a large and symptomatic tumor was not in the patient's best interest, and neither radiosurgery nor radiation therapy as sole treatments was ideal for that tumor volume and location. When discussing surgical options, aggressive versus tailored, we recommended the latter, because it would most

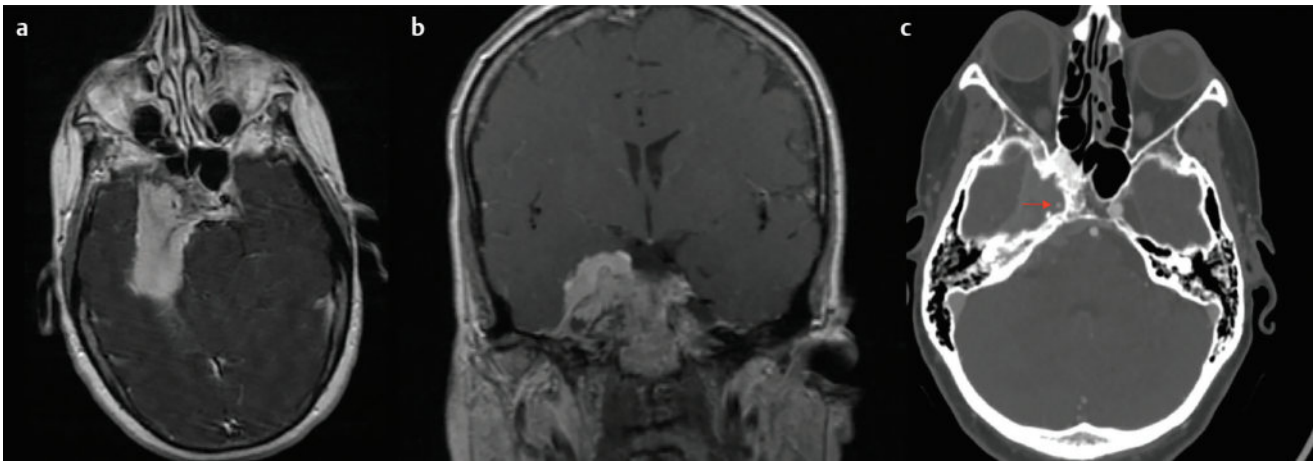


Fig. 6.1 Preoperative images. (a, b) Imaging showed an extensive skull base lesion, which was homogeneously enhancing and involving the cavernous sinus on the right, as well as the tentorium, extending into the medial middle fossa floor, Meckel's cave, the crural cistern, ambient cistern, and the lateral prepontine cistern in the posterior fossa. **(c)** A CT angiogram showed that the carotid artery was encircled by the tumor and attenuated (*arrow*).

likely reverse the patient's cranial neuropathies in the medium term (3–6 months), and result in effective long-term tumor control, particularly if the residual is treated with radiosurgery, our preferred radiation modality for small tumor volumes. The possibility of a vascular injury and stroke, temporal lobe contusion or hemorrhage, worsening of the patient's cranial neuropathies, and cerebrospinal fluid (CSF) leak and wound-related complications were discussed at length before an informed consent was signed. The patient was specifically told that her diplopia will get worse after surgery, improving 3 to 6 months later.

Though stopping short of a total resection, an option that would likely cause permanent neuropathies leading to an unusable, closed right eye, the "tailored" resection we proposed is still rather aggressive with intended residual only in the cavernous sinus. A lesser degree of resection is yet another option, but this would decrease the chances of a good functional and oncological outcome.

■ Anatomical and Therapeutic Considerations

The patient's skull base meningioma was anatomically complicated, involving multiple critical anatomical structures and compartments. The patient's symptoms were secondary to compression of the abducens nerve, either within Dorello's canal, or within the cavernous sinus, and compression of the oculomotor nerve, either within the oculomotor cistern, or within the cavernous sinus proper. The tumor involved the tentorium from the base of the anterior clinoid, to just beyond the level of the quadrigeminal plate posteriorly. It also extended laterally into the middle fossa floor, abutting the mesial temporal lobe, inferiorly into the ambient and lateral prepontine cisterns, as well as superiorly into the lateral crural cistern, the opticocarotid cistern, and the posterior roof of the cavernous sinus, where it involved the oculomotor triangle. In addition, the tumor extended into the cavernous sinus, and occupied all its compartments on the right side, with narrowing of the lumen of the cavernous segment of the carotid artery (**Video 6.1**).

Meticulous knowledge of the surgical anatomy of the cavernous sinus and surrounding structures is essential for any operation on this complex tumor. The roof of the cavernous sinus has an anterior triangle, also known as the clinoidal triangle, which is covered by the anterior clinoid process and contains the clinoidal segment of the ICA medially and the oculomotor nerve laterally. An anterior clinoidectomy is key to unlock and access the roof of the cavernous sinus, exposes the anterior component of the oculomotor nerve within the cavernous sinus and the clinoidal ICA, and allows mobilization and decompression of the intracanalicular optic nerve. The posterior triangle of the roof of the cavernous sinus, the oculomotor triangle, is bound by the anterior and posterior petroclinoid and the interclinoid ligaments, and it is penetrated by the oculomotor nerve that travels above the roof of the cavernous sinus but below the anterior petroclinoid dural fold before it truly enters the cavernous sinus at the clinoidal triangle (**Fig. 6.2**).

The surgical goal of decompressing the cranial nerves, and maximally removing the extracavernous component of the tumor, can only be achieved with relative safety by following two main principles: one, the cavernous sinus should be "unlocked" by completing a physical separation of the cavernous sinus proper from the meningeal layer of the dura that "locks" it; two, the cranial nerves and the carotid should be identified outside the cavernous sinus, and then followed into it. This requires an operation done in two interconnected surgical stages: extradural and intradural.

Fig. 6.3 briefly reviews some relevant surgical anatomy, while **Fig. 6.4** shows the extra- and intradural modular components of the planned procedure, which are named after their iconic inventors. The "Hakuba technique" refers to the mobilization of the temporal lobe dura away from the lateral wall of the cavernous sinus, V2 and V3 from anterior to posterior, for which identification and transection of the meningo-orbital dural fold is key. The "Kawase technique" refers to a posterior-to-anterior middle fossa dura dissection to expose the petrous apex, which has the greater superficial petrosal nerve (GSPN) as a key landmark. The "Dolenc technique" is performed intradurally, and refers to the division of the anterior petroclinoid fold at the oculomotor triangle, which liberates the oculomotor nerve and

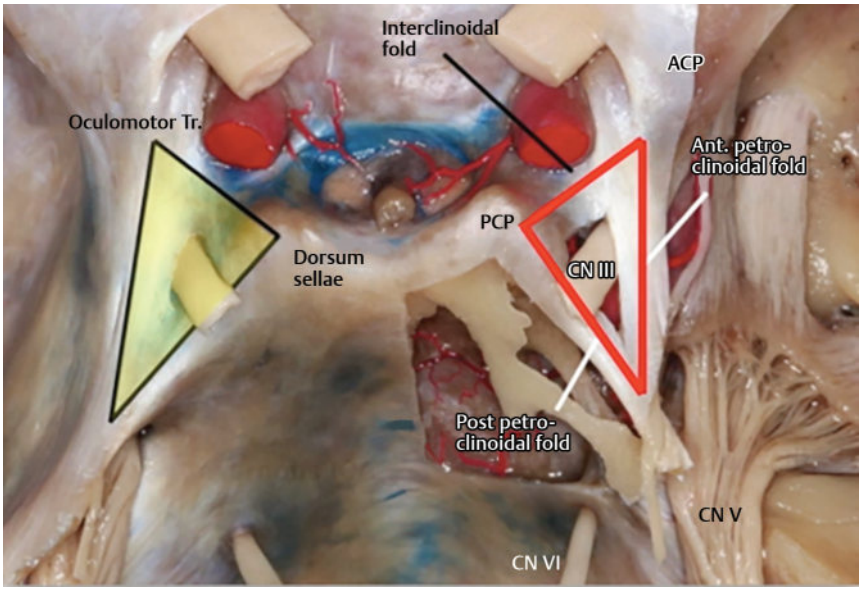


Fig. 6.2 The anatomy of the oculomotor triangle. The oculomotor triangle forms the posterior roof of the cavernous sinus, and is bordered by the anterior petroclinoid fold or ligament, the posterior petroclinoid fold, and the interclinoid fold. ACP, anterior clinoid process; PCP, posterior clinoid process; CN, cranial nerve.

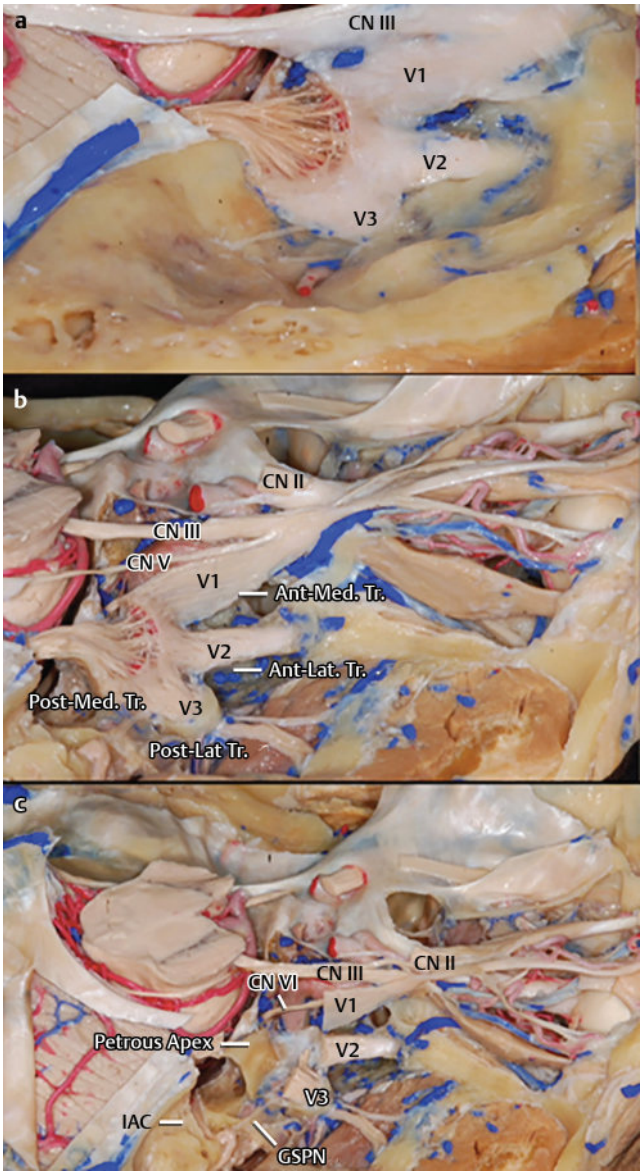


Fig. 6.3 Anatomy of the cavernous sinus and middle fossa. (a) After peeling the meningeal dura, the cavernous sinus and Meckel's cave are exposed in the middle fossa floor. **(b)** Removal of the dura of the cavernous sinus exposes the nerves in its lateral wall. The four middle fossa triangles are depicted (anteromedial, anterolateral, posteromedial, and posterolateral). **(c)** Meckel's cave is removed to show the lacerum foramen and posterior compartment of the cavernous sinus, and the petrous apex is removed posteromedial to the greater superficial petrosal nerve (GSPN) to expose the posterior fossa. CN, cranial nerve; IAC, internal acoustic canal.

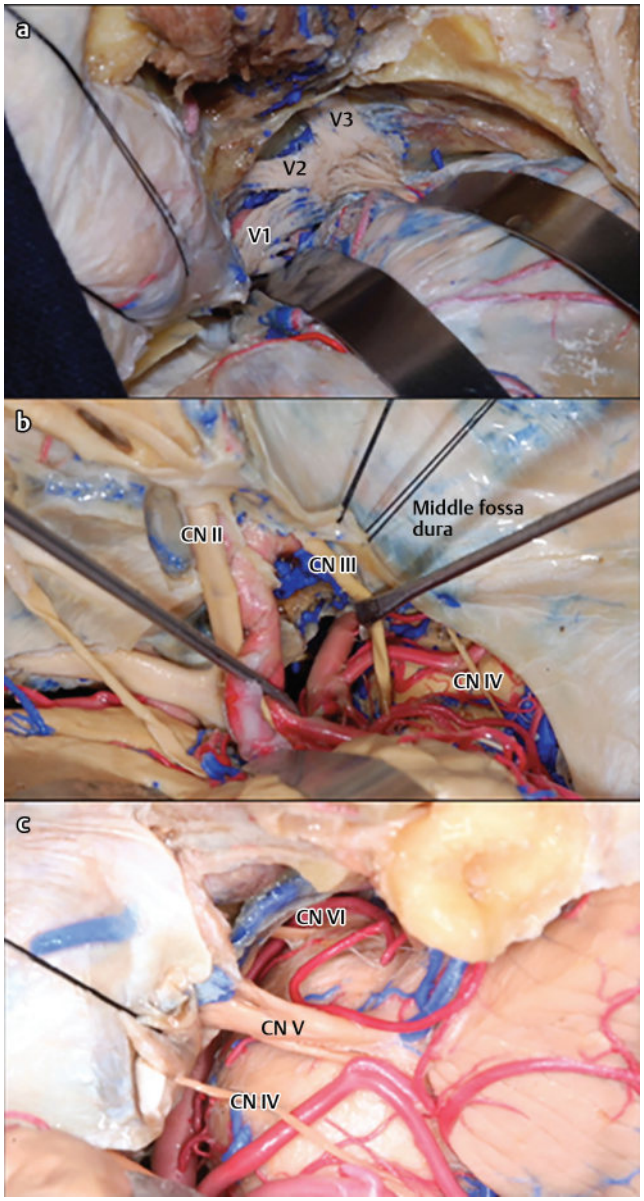


Fig. 6.4 The Hakuba–Dolenc–Kawase technique. (a) The Hakuba technique refers to the mobilization of the temporal lobe dura away from the wall of the cavernous sinus, V2 and V3 from anterior to posterior. **(b)** The Dolenc technique is performed intradurally, and refers to the division of the anterior petroclinoid fold at the oculomotor triangle, with subsequent dissection of the temporal dura from superior to inferior. **(c)** The Kawase technique refers to a posterior-to-anterior dural dissection, which exposes the petrous apex, medial to the greater superficial petrosal nerve. CN, cranial nerve.

allows subsequent dissection of the temporal dura away from the cavernous sinus from superior to inferior.

Options for Approach

While considering the craniotomy to accomplish the tumor resection and decompression of the cavernous sinus, an orbitozygomatic (OZ) craniotomy would provide excellent exposure, because it helps to minimize the manipulation of the frontal lobe (orbital osteotomy) and the temporal lobe (zygomatic osteotomy). Alternatively, a simple pterional craniotomy might be considered, but it would be less effective in mobilizing the frontal and temporal lobes. Likewise, a "minimally invasive" lateral orbitotomy through a lateral canthal incision might provide good middle fossa exposure but not quite enough access for petrosectomy, anterior clinoidectomy, and tentorial resection.

Approach of Choice

We chose to use an OZ craniotomy, which would be followed by an extradural extended middle fossa approach with an anterior petrosectomy and an anterior clinoidectomy, utilizing a combination of the Hakuba, Dolenc, and Kawase's techniques to unlock the cavernous sinus from the meningeal layer of dura.

Several critical anatomical structures are at risk during this approach. These include the optic nerve and clinoidal carotid artery during the anterior clinoidectomy, the oculomotor nerve during the dissection of the roof of the cavernous sinus, the trochlear nerve during the resection of the tentorium, the trigeminal nerve during the anterior petrosectomy or the opening of the posterior fossa dura, the abducens nerve at Dorello's canal, and finally, the superior cerebellar artery (SCA), posterior cerebral artery, basilar artery, or carotid artery.

Questions

1. What is the meningo-orbital band, and what is its significance in separating the meningeal dura from the cavernous sinus dura?
2. Other than the orbitotomy, what else can increase the safety of mobilizing the temporal lobe?
3. What cranial nerve would be at highest risk for injury during this procedure?

Description of the Technique

After endotracheal intubation, steroids (dexamethasone 10 mg), antiepileptics (Levetiracetam 1,000 mg), antibiotics (Cefazolin 2 g), and mannitol (25 g) were administered. A lumbar drain was placed at the beginning of the procedure to facilitate CSF drainage and brain relaxation. Then, the patient was positioned supine with an ipsilateral shoulder roll, and with the head in three-pin fixation. The head was rotated toward the opposite side approximately 30 to 45 degrees. It is important to avoid rotating the head more than 45 degrees because the temporal lobe will have to be manipulated more to open the sylvian fissure. Dorsal extension was used so that the malar eminence was positioned at the top of the surgical field; this helps with separating the frontal lobe from the anterior skull base floor. Lateral extension also assists in separating the temporal lobe from the middle fossa floor. The abdomen was prepped for fat graft (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: anterolateral

Craniotomy: orbitozygomatic

Modifiers: anterior clinoidectomy, anterior petrosectomy, mobilization of the cavernous sinus lateral wall

A frontotemporal incision was made and the scalp flap was elevated using an *interfascial* dissection technique to preserve the frontalis branch of the facial nerve (in which the plane of dissection is developed between the superficial and deep layers of the temporal fascia as described originally by Yaşargil). The benefit of the interfacial versus the subfascial technique is that it provides a natural plane of dissection that will expose the zygomatic arch completely without any need for fascial incisions. The temporalis muscle was encircled posteriorly to avoid cutting through muscle fibers, and the temporal fascia was incised and detached from the zygomatic process of the frontal bone, superior temporal line, and supramastoid crest. Blunt dissection is employed to elevate the deep (periosteal) temporal fascia away from the bone, avoiding electrocautery at all times to preserve the vascular supply to the muscle, which enters the deep temporal fascia from the infratemporal fossa (middle and deep temporal arteries). Once the muscle was completely detached, the only remaining attachments will be at the coronoid process and the infratemporal crest; this will allow for total rotation of the muscle inferiorly. At this point, a frontotemporal craniotomy was performed; then, the temporal muscle was reflected upward to complete the orbitozygomatic osteotomy separately, the key steps of which are summarized in **Fig. 6.5** and **Fig. 6.6**.

Extradural Dissection of the Middle Fossa

Subsequently, we proceeded with the extradural portion of the procedure. We started with dissection of the middle fossa dura from the true lateral wall of the cavernous sinus, in an *anterior-to-posterior* direction, as first described by Hakuba. The first step in this process involves the coagulation and

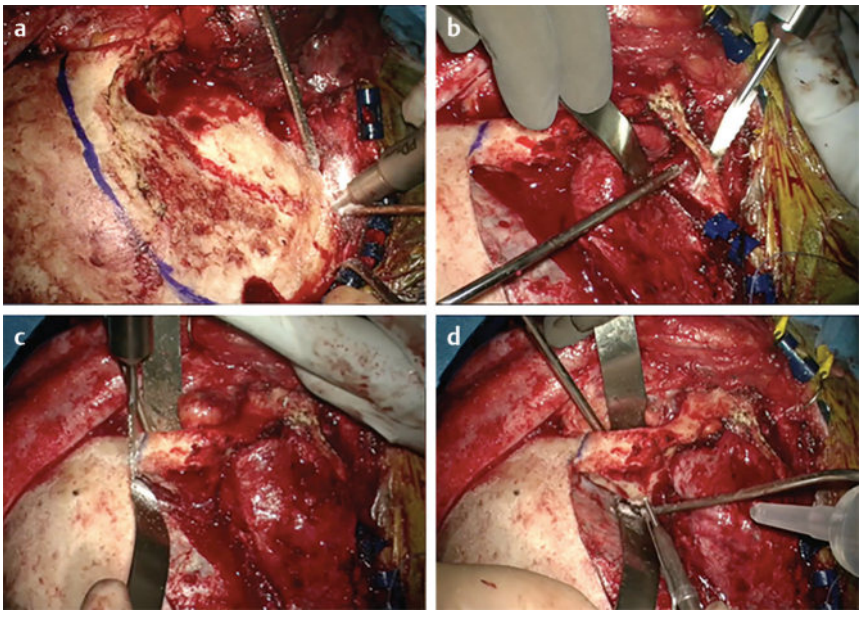


Fig. 6.5 Steps of orbitozygomatic craniotomy. (a) The craniotomy is performed first using three burr holes. Subsequently osteotomies are performed at **(b)** the root of the zygoma, **(c)** the orbital rim, **(d)** and the roof of the orbit.

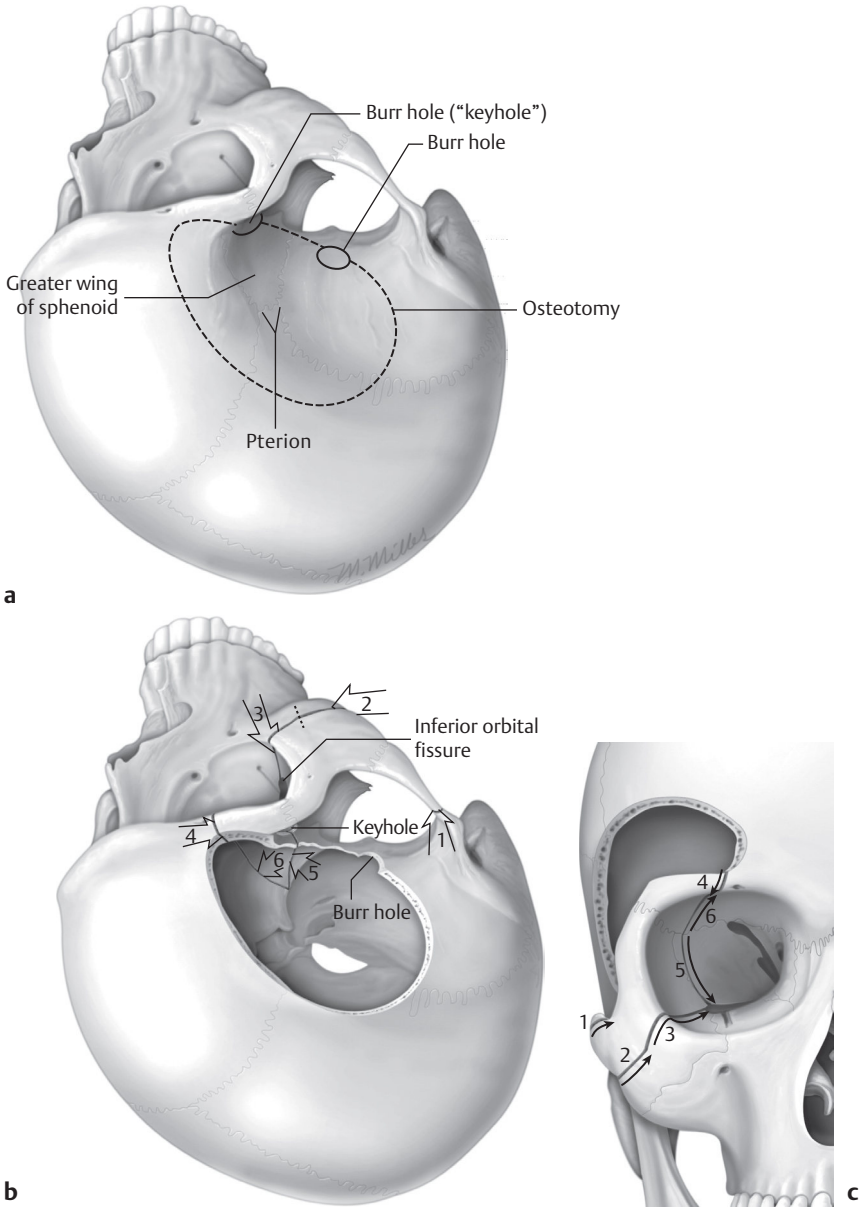


Fig. 6.6 Two-piece orbitozygomatic craniotomy. (a) Extent of a first-stage craniotomy in the standard frontotemporal location. **(b)** Surgeon's view showing osteotomies necessary for removal of orbitozygomatic flap. **(c)** Frontal view of the same. The cuts are made is this order: 1, root of the zygoma; 2, inferior zygoma; 3, lateral orbit; 4, superior orbit; 5, inferior orbital fissure to sphenoid wing; 6, sphenoid wing to superior orbit. (Reproduced from Friedman R, Slattery III W, Brackmann D, et al., eds. Lateral Skull Base Surgery. The House Clinic Atlas. 1st ed. Thieme; 2012.)

division of the meningo-orbital fold, which "unlocks" the temporal dura from the orbit (the meningo-orbital fold is a dural connection of the anterior temporal dura with the periorbita, which transmits the recurrent lacrimal artery—an anastomotic vessel connecting with branches of the middle meningeal artery). The temporal dura was then peeled away from the cavernous sinus wall using a combination of sharp and blunt dissection, exposing from superior to inferior: the anterior clinoid, superior orbital fissure, maxillary strut, and V2. The dura of the middle fossa floor was further dissected to expose the anterior aspect of V3 and Meckel's cave, and this was key to identifying the middle meningeal artery at the foramen spinosum. It is the preference of the senior author (JFM) to drill around the foramen spinosum to open it and coagulate the artery within the foramen rather than above it, as this avoids unnecessary bleeding, excessive coagulation, and potential dural breaches at the middle fossa dura.

The dissection of the temporal dura was then performed in a *posterior-to-anterior* fashion, as described by Kawase, to avoid traction on the GSPN and geniculate ganglion (facial nerve), and to define the boundaries of the anterior transpetrosal approach: the GSPN, the arcuate eminence, the superior petrosal sinus at the petrosal ridge, and the posterior border of V3. It is important to follow the GSPN underneath V3, as it is at this most anterior aspect of the petrous apex where the petrous carotid canal is typically thinnest, allowing identification of the petrous ICA as it enters the foramen lacerum. The Doppler ultrasound can assist in confirming the location of the ICA, and with monopolar electrical stimulation we can confirm the trajectory of the GSPN. It is also important to remember that in approximately 15% of cases, the geniculate ganglion is dehiscent and susceptible to iatrogenic injury. The steps are summarized in **Fig. 6.7**.

Drilling of the Skull Base

The bone of the anteromedial triangle of the middle fossa, overlying the superior orbital fissure and V2, was involved with

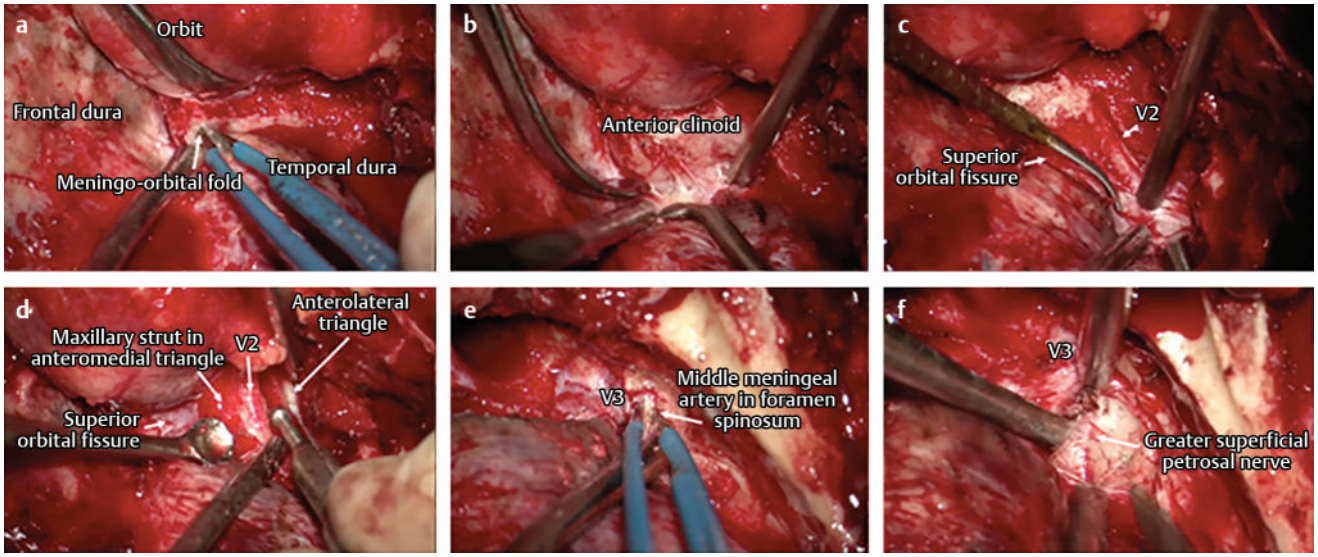


Fig. 6.7 Temporal lobe dura. Steps of detachment of temporal dura from the true wall of the cavernous sinus from anterior to posterior (the Hakuba technique), and posterior to anterior (the Kawase technique). **(a)** After coagulation and division of the meningo-orbital fold, the temporal dura is unlocked and can be detached from the lateral wall of the cavernous sinus **(b, c)**. Some of the sphenoid wing is drilled to skeletonize the superior orbital fissure and V2, as well as to create space in the anterolateral triangle to allow anterior displacement of V3 **(d)**. **(e)** Coagulation and division of the middle meningeal artery allows a posterior-to-anterior detachment of the temporal dura **(f)**.

tumor and was drilled extensively with caution not to enter the lateral recess of the sphenoid sinus. The anterolateral triangle was also drilled to allow for anterior mobilization of V3, but also avoiding unnecessary exposure of the infratemporal fossa. Before the petrosectomy we identified the approximate position of the internal acoustic canal (IAC) (approximately parallel to a line bisecting the angle of the GSPN, and the arcuate eminence, while confirming with image guidance), and the estimated location of the cochlea (at the intersection between IAC and GSPN). Then, we identified the trajectory of the petrosal ICA segment as explained above, by identifying dehiscent bone underneath V3 and at the lacerum foramen, and confirmed it with image guidance and Doppler ultrasound. We then started drilling at the most medial area of the petrous apex, where it is safest, and proceeded from medial to lateral and from posterior to anterior. The lateral limit is the cortical bone of the petrous ICA canal, or the petrous ICA itself, the posterolateral limit is the cochlea, the posteromedial limit is the dura of the IAC, and the inferior (deep) limit is the inferior petrosal sinus and petroclival fissure. It is key to realize that the anterior limit of the anterior petrosectomy is not V3, but the foramen lacerum, but it is very difficult to access the foramen lacerum without full mobilization or transection of V3. After completing the petrosectomy and exposing the dura of the posterior fossa, a small durotomy allows for direct access to the lateral cerebellomesencephalic cistern for CSF egress and further brain relaxation (**Fig. 6.8**).

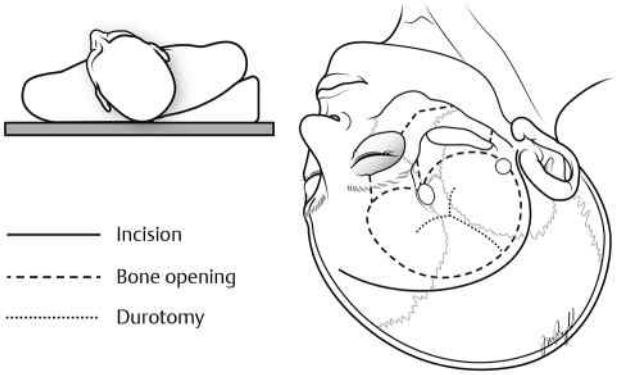
Operative Setup

Position: supine, head turned 30 degrees

Incision: root of zygoma to contralateral mid-pupillary line

Bone Opening: frontotemporal orbitozygomatic

Durotomy: T-shaped, with long axis along the sylvian fissure; temporal limb into the posterior fossa



We then proceeded with an anterior clinoidectomy performed in an extradural fashion. We began the clinoidectomy by identifying the optic nerve extradurally as it entered the optic canal. The roof of the canal was removed using precise high-speed drilling with constant irrigation. Next, we sequentially "debulked" the body of the clinoid process and detached it from its three attachments—the planum sphenoidale (optic canal) superiorly, the sphenoid lesser wing laterally, and the optic strut inferior to the optic nerve in the canal (**Fig. 6.8c–e**). Finally, the remaining clinoid was carefully dissected from its dural attachments and removed completely to expose below the clinoidal triangle of the cavernous sinus roof, with the paraclinoid ICA medially and the oculomotor nerve laterally (see **Operative Setup).**

Intradural Operation

After completing the extradural part of the operation, the dura was opened in a T-shaped fashion with the long axis parallel to the sylvian fissure and directing the incision toward the roof of the cavernous sinus (clinoidal triangle). Stopping approximately

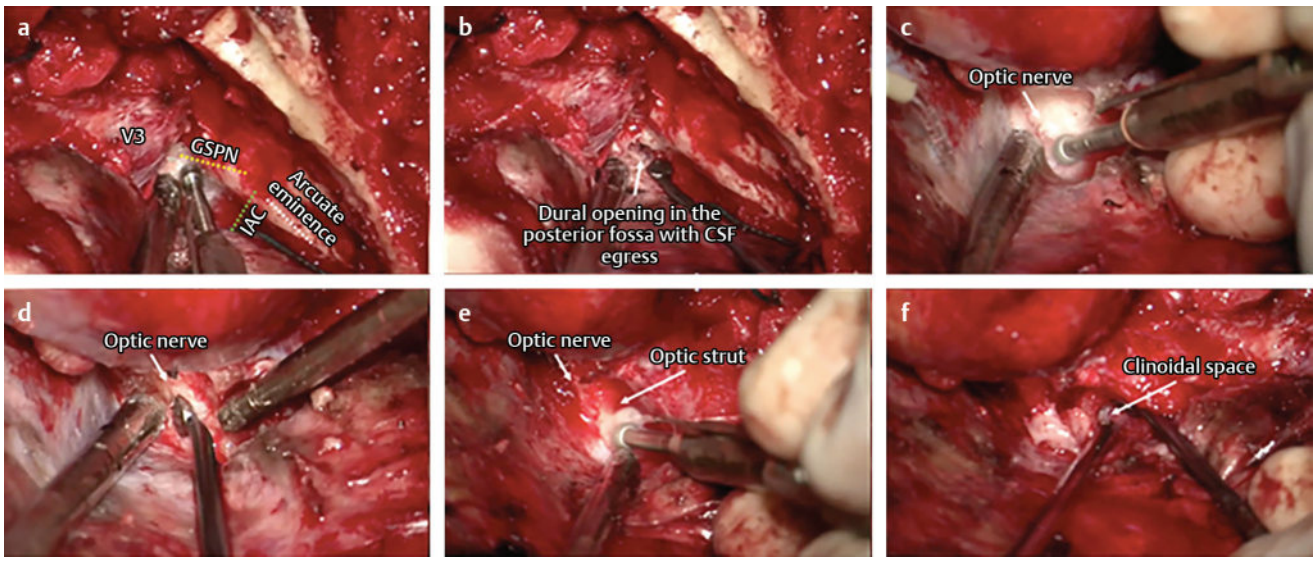


Fig. 6.8 Skull base drilling. (a) A line bisecting the axis of the arcuate eminence and the greater superficial petrosal nerve (GSPN) approximates the location and trajectory of the internal acoustic canal (IAC). **(b)** An opening in the posterior fossa dura releases CSF and affords brain relaxation. **(c)** The anterior clinoidectomy starts by drilling the roof of the optic canal to an eggshell thickness of bone, which is subsequently easily removed **(d)**. **(e)** Drilling of the optic strut allows disconnection of the anterior clinoid. After the three attachments of the anterior clinoid are detached (planum, sphenoid ridge, and optic strut), the remainder of the clinoid is removed **(f)** exposing the clinoidal space, which contains the clinoidal segment of the carotid artery at the depth.

1 cm distal to the cavernous sinus and optic canal, the dura was then cut parallel to the superior orbital fissure inferiorly (temporal side), and parallel to the falciform ligament (frontal side). This type of incision facilitates simultaneous extra- and intradural access, and it is designed to remove the temporal lobe dura infiltrated with tumor.

The sylvian fissure was opened in a typical inside-out technique up to the basal cisterns, and with continuous arachnoid dissection the oculomotor nerve was identified and dissected from the uncus laterally and the PCOM (posterior communicating artery) and ICA medially, and followed from the interpeduncular fossa and cerebral peduncle down to the oculomotor triangle, where it was encased by tumor (**Fig. 6.9**). Opening Liliequist's membrane and freeing the arachnoid adhesions around the oculomotor nerve allowed further mobilization of the temporal lobe. After resecting the tumor occupying the parapeduncular space around the oculomotor nerve, the oculomotor triangle was clearly defined, and we proceeded with incising the *anterior petroclinoid ligament*. This allowed further peeling of the temporal dura, now in a *superior-to-inferior fashion*, as first described by Dolenc (**Fig. 6.10**). This critical step joined the extradural with the intradural components of the operation, completely "unlocking" the true wall of the cavernous sinus from all its dural attachments, and allowed for decompression of the oculomotor nerve that was being strangulated by tumor growing within the cavernous sinus and pushing the nerve against the anterior petroclinoid ligament. Then, the separation of the middle fossa dura from the cavernous sinus was completed, progressing from superior and anterior to inferior and posterior, leaving cranial nerves III, IV, and V1 with the cavernous sinus. Then, a posterior dural cut, completed just anterior to the entrance of cranial nerve IV into the cavernous sinus, allowed the complete resection of the temporal dura. This dural cut only involved the temporal (meningeal) dura that has already been dissected away from the cavernous sinus and the cranial nerves inside.

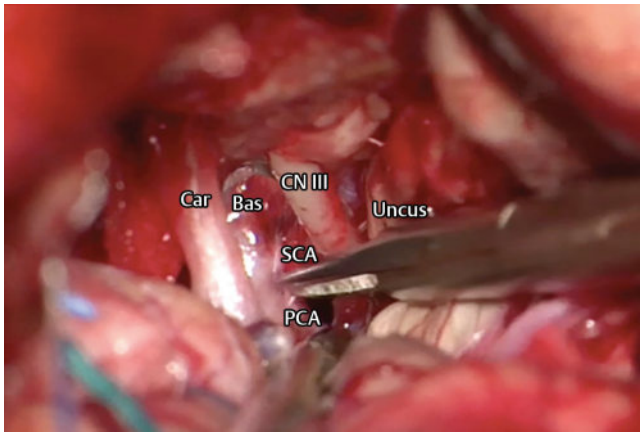


Fig. 6.9 Intradural steps. Beginning the intradural component of the operation by splitting the sylvian fissure, and skeletonizing the oculomotor nerve, freeing it from all the arachnoid adhesions. The basilar artery (Bas), posterior cerebellar artery (PCA), and superior cerebellar artery (SCA) are all visible between the carotid (Car), and the oculomotor nerve (CN III).

Once the temporal dura was resected, we then focused our attention on resecting the tentorium that was enlarged and invaded by tumor, as well as resection of the tumor within the infratentorial compartment. The trigeminal root was identified at the lateral prepontine cistern, freed from tumor, and followed to the porus trigeminus, where again it was encased by tumor. The dural ring along the porus trigeminus was opened to facilitate tumor resection within Meckel's cave, but also to allow further mobilization of the trigeminal nerve and the tentorium, and widening of the access to the infratentorial space. The tentorium was then transected from lateral to medial, while taking care to identify and preserve the trochlear

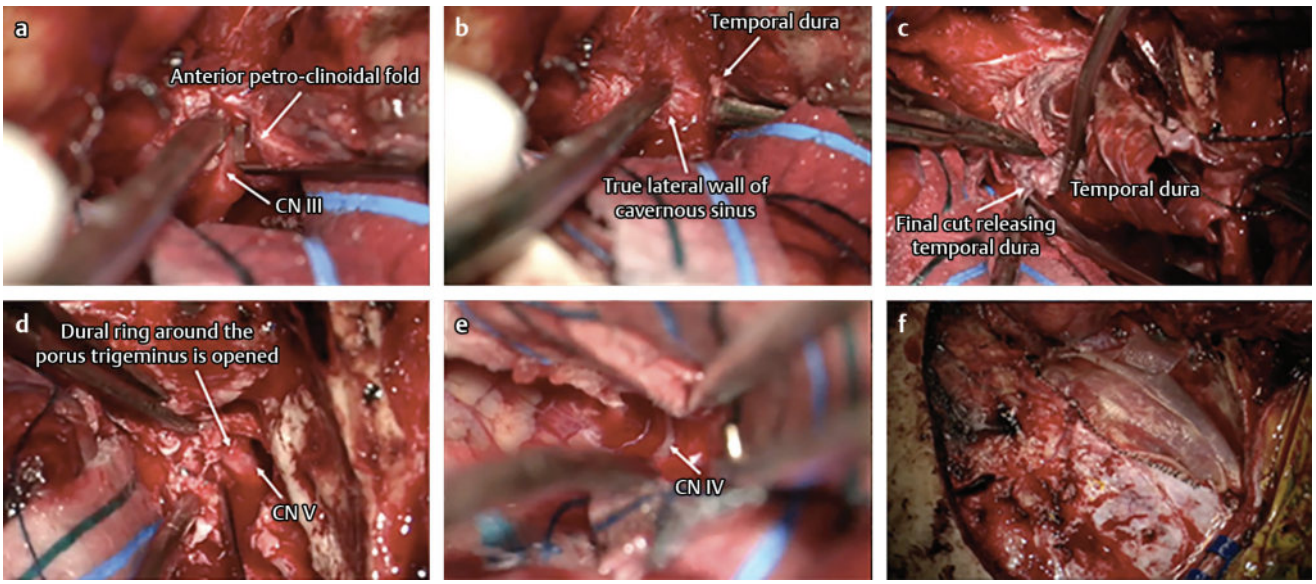


Fig. 6.10 Key steps in resecting the temporal dura, and the tentorium. **(a)** The anterior petroclinoid ligament, which constitutes the lateral border of the oculomotor triangle, is incised along the oculomotor nerve. **(b)** This allows the detachment of the temporal dura from the true wall of the cavernous sinus from superior to inferior, as described by Dolenc, constituting the final step in "unlocking" the cavernous sinus from all its dural attachments. **(c)** The temporal dura can now be resected. We then proceed with resection of the tentorium. **(d, e)** The dural ring around the porus trigeminus is opened to allow further tumor resection, and finally the tentorium can be completely resected, taking care not to injure CN IV. **(f)** Finally, the dural defect is repaired with a dural substitute.

nerve as well as the posterior cerebral, and the SCAs and their branches. The subtemporal intradural route was used to identify the trochlear nerve entering the cavernous sinus just before completing the most medial tentorial cut, and the tentorium was transected just behind this entry point. The superior petrosal sinus was barely patent in this case, given the extensive tumor involvement. The tentorium was resected as far posterior as the quadrigeminal cistern and posterior incisural space. While resecting the tentorium, a tentorial branch of the SCA was identified and unfortunately avulsed before selective coagulation. The SCA was repaired with clip reconstruction and preservation of the vessel (**Fig. 6.11**).

At the conclusion of the resection stage, we had obtained resection of all tumor components except inside the cavernous sinus and in Dorello's canal. Cranial nerves III to VI had been identified and preserved anatomically (**Video 6.2**).

Surgical Pearls

1. A critical surgical concept is "unlocking" the true wall of the cavernous sinus from its surrounding dura. This requires a combined extra- and intradural approach, using the techniques described by Hakuba, Kawase, and Dolenc, and a combined extra- and intradural tumor resection, which allows the surgeon to maximize and compartmentalize the resection of the tumor while preserving key neurovascular structures.
2. Identification of the cranial nerves outside the cavernous sinus, where they are not involved with tumor, maximizes the chances of their preservation and decompression. This allows the surgeon to follow them into their cavernous sinus entry point to separate the extracavernous tumor compartment from the intracavernous compartment where the nerves are located.
3. Every successful skull base resection starts with proper planning and approach selection, and every step of the operation should serve a clear purpose. In this case, the anterior clinoidectomy was key to access the cavernous sinus roof, transect the anterior petroclinoid ligament, and allow resection of middle fossa dura with preservation of oculomotor nerve; the anterior petrosectomy was key to identify the trigeminal nerve, access the infratentorial space, and open Meckel's cave; the sylvian fissure was open to mobilize the temporal lobe and to find the cisternal segment of the oculomotor nerve; the subtemporal intradural approach was needed to identify the cisternal trochlear nerve and transect the tentorium.
4. Dedicated training and study in the surgical neuroanatomy laboratory is paramount to becoming comfortable applying complex skull base procedures. Presurgical planning with precise imaging studies (CT angiography/CT venography, fine-cut MRI with FIESTA sequences) is important and newer three-dimensional printing and virtual reality simulations are interesting additions, but comprehensive understanding of the complex three-dimensional microsurgical anatomy should precede all of them.

■ Aftercare

The lumbar drain was kept at the completion of the procedure, and the patient was drained at 5 to 10 cc/h for 72 hours to prevent pseudomeningocele formation. An immediate postoperative MRI showed complete resection of all but the cavernous component of the tumor (**Fig. 6.12**). The patient did well clinically after her surgery, but had complete cranial nerves III and IV, and partial VI palsies immediately postoperatively. She was discharged home on postoperative day 5, and at 3 months' follow-up had a

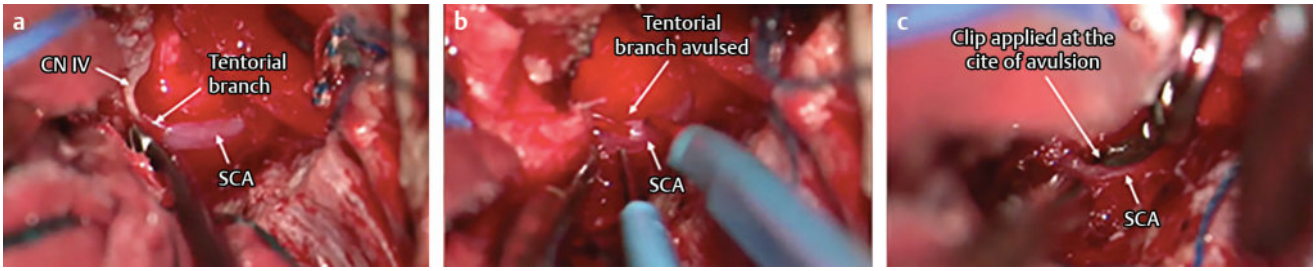


Fig. 6.11 Vascular injury. During the resection of the tentorium **(a)**, a tentorial branch of the superior cerebellar artery (SCA) was avulsed **(b)**. This was repaired with an angled clip on the side of the SCA, which was not occluding the primary vessel **(c)**.

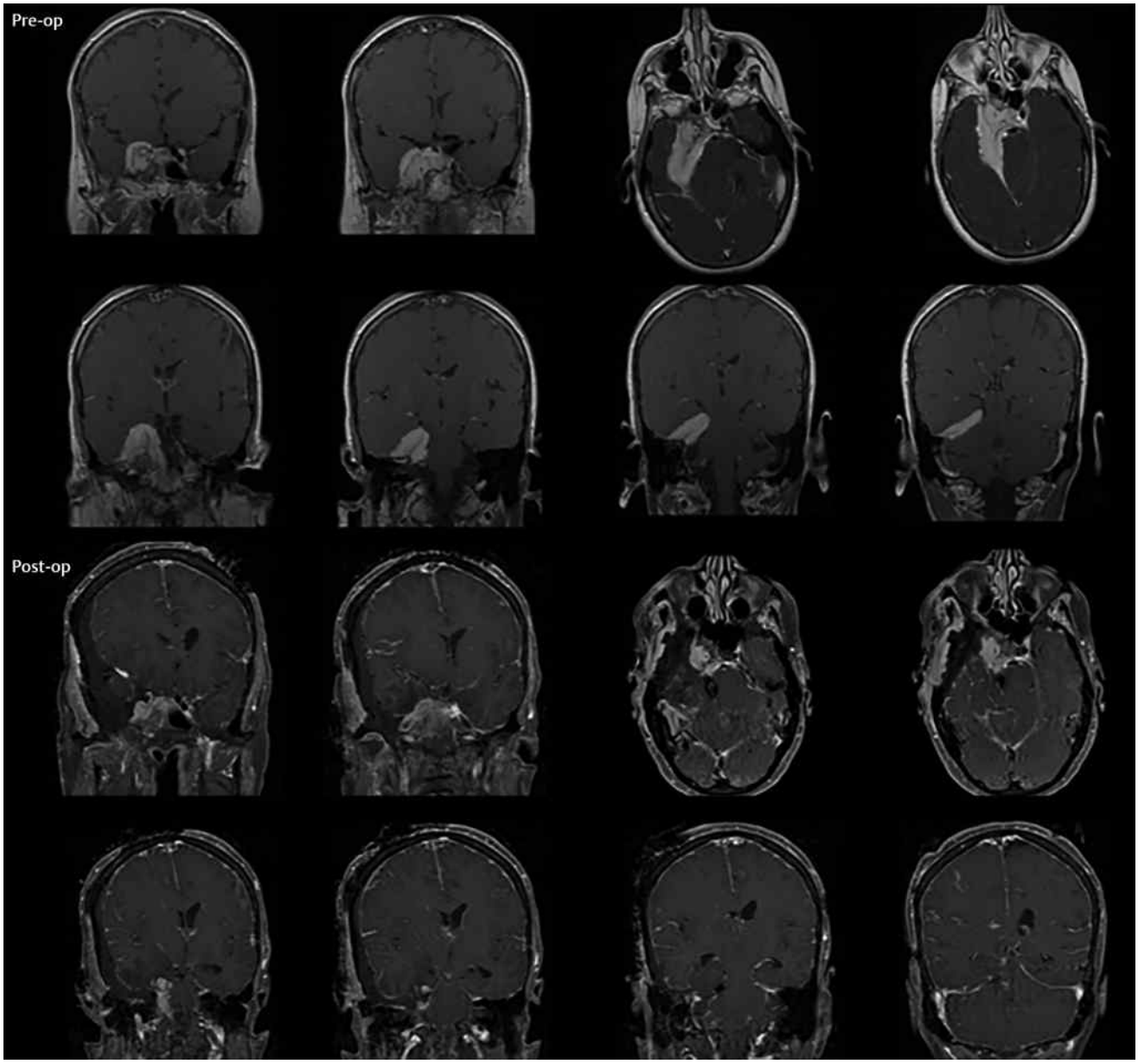


Fig. 6.12 Surgical result. Comparison of the preoperative and postoperative imaging showing complete resection of all but the cavernous sinus components of the tumor.

near-complete resolution of all her palsies, except a mild partial VI palsy. The result of histopathological analysis was consistent with a grade I meningioma. After discussion with the patient, she underwent radiosurgery to the remaining cavernous sinus component of the tumor. At 6 months' follow-up the abducens palsy had completely resolved, and the patient had normal extraocular movements with no diplopia.

■ Possible Complications and Associated Management

Vascular injuries are potentially devastating in these extensive procedures, and the most feared complication. For this reason, a CT angiogram is obtained in all skull base procedures in our practice to study the vascular relationship of the lesion. Occasionally, a formal angiogram may be indicated, particularly if preoperative embolization is contemplated, and an additional BTO is performed if the sacrifice of a major vessel is considered. Indeed, a complication with potentially devastating consequences happened in our case with the avulsion of a tentorial branch of the SCA. Although initially coagulation was attempted at the site of the avulsion, this was abandoned because of the risk to the parent vessel. Suturing the vessel was not a good option, given the limited surgical corridor, and the risks of postoperative pseudoaneurysm or vessel stenosis. Ultimately a small curved clip was used to reconstruct the side wall of the artery with sufficient preservation of flow in the parent vessel, as confirmed with intraoperative Doppler ultrasound. There was no evidence of postoperative stroke on diffusion imaging.

Another major category of complications is CSF leaks and pseudomeningoceles. When the middle fossa dura is extensively resected, watertight closure is impossible, simply because the dural resection has extended to the distal dural ring, optic canal dura, and cavernous sinus lateral wall. In this case, the dura was closed primarily where possible, and then reconstructed at the temporal area with a pericardial graft, which was tucked to the floor of the middle fossa. This was reinforced with an inlay collagen matrix, and onlay fat to occupy all the empty space. A small piece of fat was also used to seal the petrosectomy defect, despite the lack of open air cells, and the opening into the posterior fossa was repaired with another small collagen matrix piece and a small fat graft on top. A muscle plug was used to seal a small breach of the frontal sinus. In addition, bone wax was used where we had breached some roof of mastoid air cells in the middle fossa floor laterally.

Transient cranial nerve palsies are to be expected. Perioperative steroids can be used to mitigate that risk. When performing an OZ craniotomy, neurophysiological monitoring of the cranial nerves can be challenging, but leads can still be inserted sterilely during the time of surgery, and this can help with identification of the nerve within the tumor. More importantly, identifying the cranial nerves outside the cavernous sinus, and where they are not encased by tumor, allows the surgeon to follow them into the tumor and its preservation.

Brain contusions from brain manipulation, particularly of the temporal lobe, as well as venous strokes secondary to sacrifice of draining veins can also be troublesome complications. Some degree of brain contusion is quite common in these procedures and usually well-tolerated, but it is paramount to maximize brain relaxation by draining CSF (lumbar drain and cisternal opening). While sacrificing temporal bridging veins may be required for this procedure, it is important to study the venous variations to identify potential patterns of drainage that can increase the risk of postoperative venous stroke, such as a basal vein with anterior drainage pattern or a large sphenoparietal sinus draining posteriorly toward the petrosal sinuses via the middle fossa.

Perspective

Harry R. van Loveren, R. Tushar Jha, and Siviero Agazzi

■ Introduction

To begin at the beginning, the authors of the preceding section are correct that the diagnosis of cavernous sinus meningioma can be made with relative certainty in the vast majority of cases, and both clinical and surgical decisions can be made with the presumption of meningioma. In a few situations, however, there can be and should be uncertainty that requires special attention. Sometimes this involves a mismatch between the radiographic appearance of a benign meningioma and the clinical presentation.

Meningiomas are very slowly progressive tumors with an average growth rate of only 1 to 2 mm per year. A patient with a cavernous sinus or petrocavernous meningioma can present with slowly progressive ophthalmoplegia, with diplopia being the most common symptom. However, if a patient presents with rapidly progressive and severe retro-orbital pain or rapid onset of ocular paralysis, other diagnoses must be considered including infection (i.e., nasopharyngeal fungal or bacterial) and malignancy (i.e., nasopharyngeal carcinoma, lymphoma). Radiographic evidence of extension of tumor along the maxillary and mandibular nerves below the skull base should also give the neurosurgeon pause in decision making.

■ Case Presentation 1

A 74-year-old man with a history of multiple resections of skin cancers, including melanoma of the nose and right frontal basal cell carcinoma, presented with persistent right ophthalmoplegia and right hemifacial numbness that developed over the course of 3 weeks. Neurological examination revealed right ophthalmoplegia with a fixed pupil and complete ptosis. He had dense numbness to pinprick and light touch in the right V1, V2, and V3 distributions, and atrophy of his right temporalis and masseter muscles. MRI revealed an 11 × 7 mm enhancing mass within the right cavernous sinus extending into Meckel's cave, and anteriorly toward the orbital apex (**Fig. 6.13a–d**).

Anatomical and Therapeutic Considerations

This patient's history of several skin cancers including melanoma and basal cell carcinoma is a "cause for pause" and should lead the neurosurgeon to consider the diagnosis of metastasis rather than a meningioma. This patient's lesion was quite small and it did not cause any mass effect on the brainstem that would necessarily require tumor debulking. Therefore, establishing a diagnosis was the primary goal. This can be accomplished by

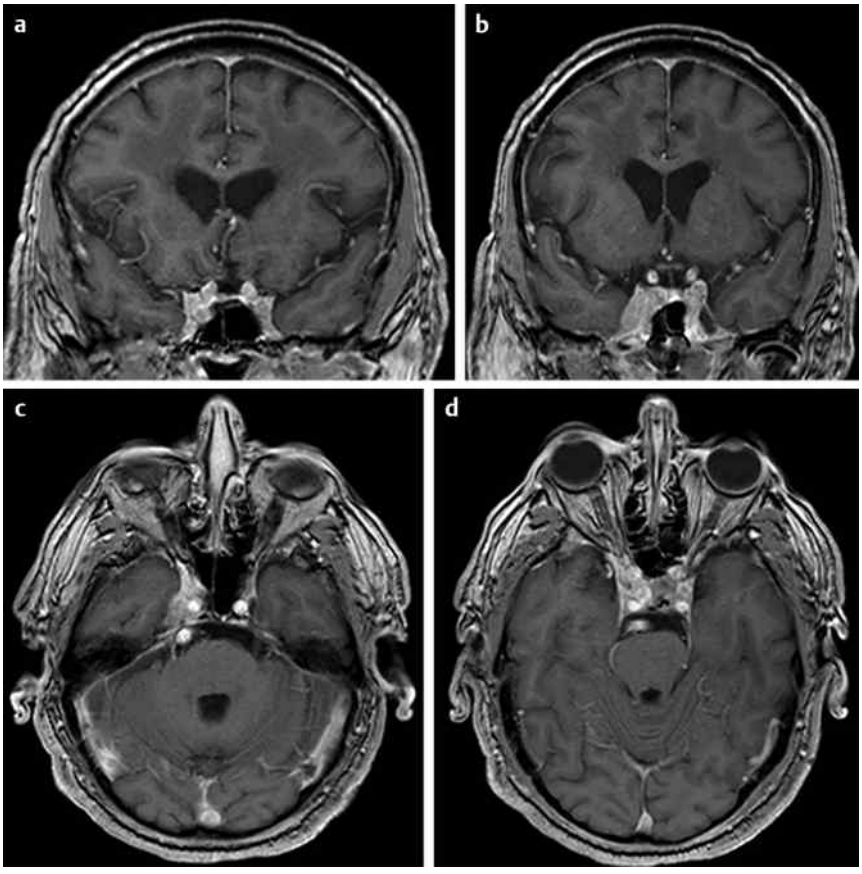


Fig. 6.13 Images of Case 1. (a, b) Coronal and **(c, d)** axial MRI with contrast showing a 11 × 7 mm enhancing mass of the right cavernous sinus extending toward the orbital apex anteriorly and Meckel's cave posteriorly. There is clearly no mass effect on the brainstem that would necessitate debulking of the tumor.

performing a biopsy, thus avoiding an extensive operation and delay in appropriate treatment.

It is essential that the "cavernous sinus surgeon" should have available strategies for biopsy, and not simply a "one-size-fits-all" operation for all surgical indications. The two techniques that we employ are endoscopic endonasal transsphenoidal biopsy of the medial cavernous sinus or the percutaneous biopsy of tumor extension into Meckel's cave through the foramen ovale—the same route used for percutaneous procedures for trigeminal neuralgia. In this patient's case, we employed the percutaneous biopsy technique to obtain tissue for diagnosis. The patient's cavernous sinus/Meckel's cave lesion was confirmed to be metastatic melanoma, and the patient was subsequently treated with chemotherapy and radiation. This case reinforces the point that the patient's medical history, clinical presentation, and differential diagnoses should be carefully considered to decide on the treatment and surgical approach. Doing so saved the patient from an extensive operation that would have failed to cure the patient, regardless of the extent of tumor resection.

When the diagnosis of meningioma is confirmed or presumed, the option of surveillance without intervention should be seriously considered. Of course, in the case presented by the previous authors, the patient was symptomatic and warranted intervention. Even then, there will be symptomatic patients for whom surgical intervention may not be wise because of questionable benefit. While considering surgery and the benefits of tumor resection, the surgeon should look carefully for the outline of the lateral dural wall of the cavernous sinus which is visible in most cavernous and sphenocavernous meningiomas. This line will distinguish what parts of the tumor are within the cavernous sinus and what is extracavernous (i.e., middle fossa, posterior fossa, etc.). The extracavernous tumor can be safely removed and the possible benefits include reduction of mass effect and pressure, decompression of regional neurovascular structures including the optic and extracavernous oculomotor and abducens nerves, and, on occasion, relief of the "compartment syndrome" that is developing in the cavernous sinus.

The management of the tumor within the cavernous sinus should be entirely different, as we discuss below.

■ Case Presentation 2

A 50-year-old woman presented to our clinic with slowly progressing right ptosis and complaints of blurry vision. She first noticed her "droopy right eye" 6 months prior to presentation. However, in retrospect she noticed it in pictures from the past 1 to 2 years. More recently, she started to complain of blurry vision specifically upon left lateral gaze. Neurological examination confirmed right eye ptosis and diplopia with left lateral gaze.

Anatomical and Therapeutic Considerations

A visit with an ophthalmologist led to an MRI of the brain. This study revealed a 3.1 × 2.7 × 2.5 cm cavernous sinus tumor (**Fig. 6.14**). Unlike the man in the previous case, this woman had no history of malignancy and had a very slow and progressive onset of symptoms, all very typical of a cavernous sinus meningioma. In her case, we can presume with great certainty that this mass was a meningioma. The lateral wall of the cavernous sinus can clearly be delineated on the MRI scans in **Fig. 6.14**. The tumor was almost completely within the cavernous sinus and thus a holocavernous meningioma.

We explained to the patient that opening the cavernous sinus to resect the meningioma posed far greater risks than benefits. It was imperative to counsel the patient that operating would in fact worsen her ophthalmoplegia in the postoperative period, and that recovery thereafter would reach no better than her preoperative baseline and not to normal. Therefore, she would most likely still have diplopia. Given the natural history of these holocavernous meningiomas and the futility of surgical resection, we elected to treat our patient with a short course of steroids and intensity-modulated radiation therapy (IMRT).

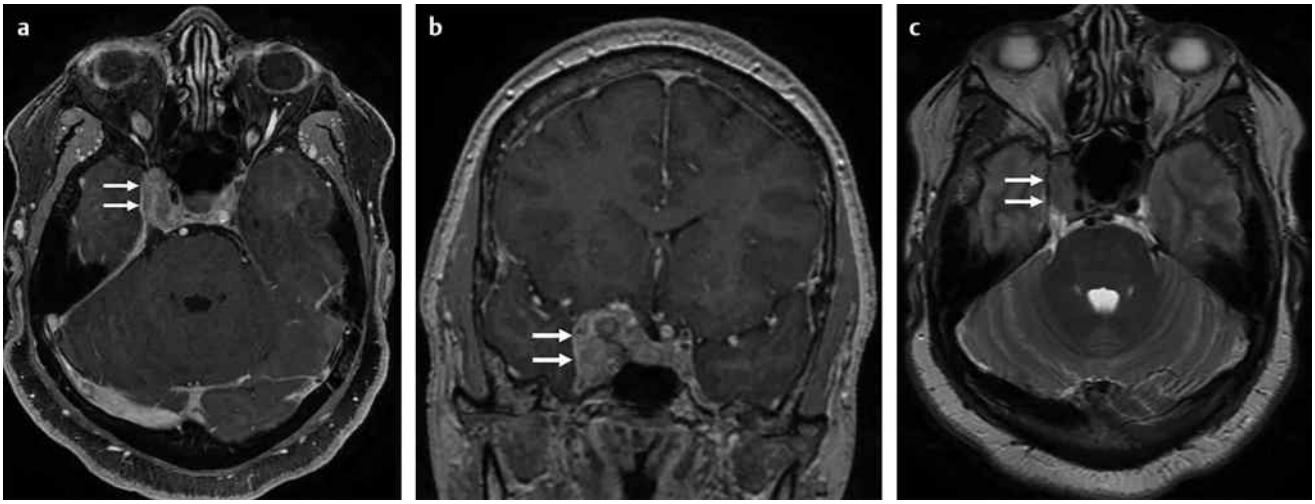


Fig. 6.14 Images of Case 2. (a) Axial and **(b)** coronal MRI brain with contrast showing an enhancing mass of the right cavernous sinus. The arrows indicate the lateral wall of the cavernous sinus. This meningioma is completely contained within the cavernous sinus. This holocavernous meningioma is best treated nonsurgically with steroids and radiation. **(c)** The arrows indicate the lateral wall of the cavernous sinus, which can be seen clearly as a dark line on this axial T2 MRI. The tumor is completely confined in the cavernous sinus.

Commentary

There is now a rather well-established sentiment among neurosurgeons that holocavernous meningioma—meningioma that fills the cavernous sinus, encases the carotid artery, without significant extension beyond the lateral wall—cannot be aggressively removed without permanent cranial nerve damage. Certainly, sacrifice and bypass of the cavernous segment of the ICA for radical benign tumor resection has been largely abandoned. Within the current "boom" in endoscopic neurosurgery, there is much discussion of performing an endoscopic endonasal transsphenoidal approach to attack these intracavernous meningioma from a medial perspective. In theory, this approach will avoid the oculomotor, trochlear, V1 and V2, which are located in the lateral aspect of the cavernous sinus and block the access into the cavernous sinus during transcranial approaches. The hope is that by avoiding these cranial nerves, resolution of ocular deficits is more likely. Although we have found this approach useful for the relief of pain, biopsy confirmation of diagnosis, and partial tumor debulking, we have had limited ability to reverse cranial nerve deficit with this technique for patients with intracavernous meningioma.

The natural history of holocavernous meningiomas has recently been reported on 53 patients that were followed for an average of 10.8 years. These tumors are indolent in nature. Furthermore, a patient's clinical symptoms were found to be independent of the meningiomas' initial size or progressive growth. The results also showed that symptomatic patients should first be treated with a 1- to 2-month course of corticosteroids, and even a second course if symptoms recur. Prisms should also be considered to alleviate diplopia in these patients. If symptomatic treatment fails, then radiation therapy such as stereotactic radiosurgery (SRS) and IMRT are suitable treatment modalities for the holocavernous meningioma.

■ Case Presentation 3

A 52-year-old woman was found to have a large left sphenocavernous meningioma measuring 5 × 4.6 × 4.8 cm (**Fig. 6.15a–d**). This mass was found on a PET (positron emission tomography) scan for surveillance of the patient's previously diagnosed inflammatory breast cancer. In retrospect, the patient endorsed fullness over the left frontotemporal region and some blurry vision over the previous 2 years. On neurological examination, the patient was remarkably intact except for a known central scotoma in the left eye.

Anatomical and Therapeutic Considerations

Though our patient has a history of breast cancer, we can still safely presume that her mass was a meningioma. A metastatic lesion would have reached this size at a much faster rate, and symptoms would have developed much quicker. Moreover, the

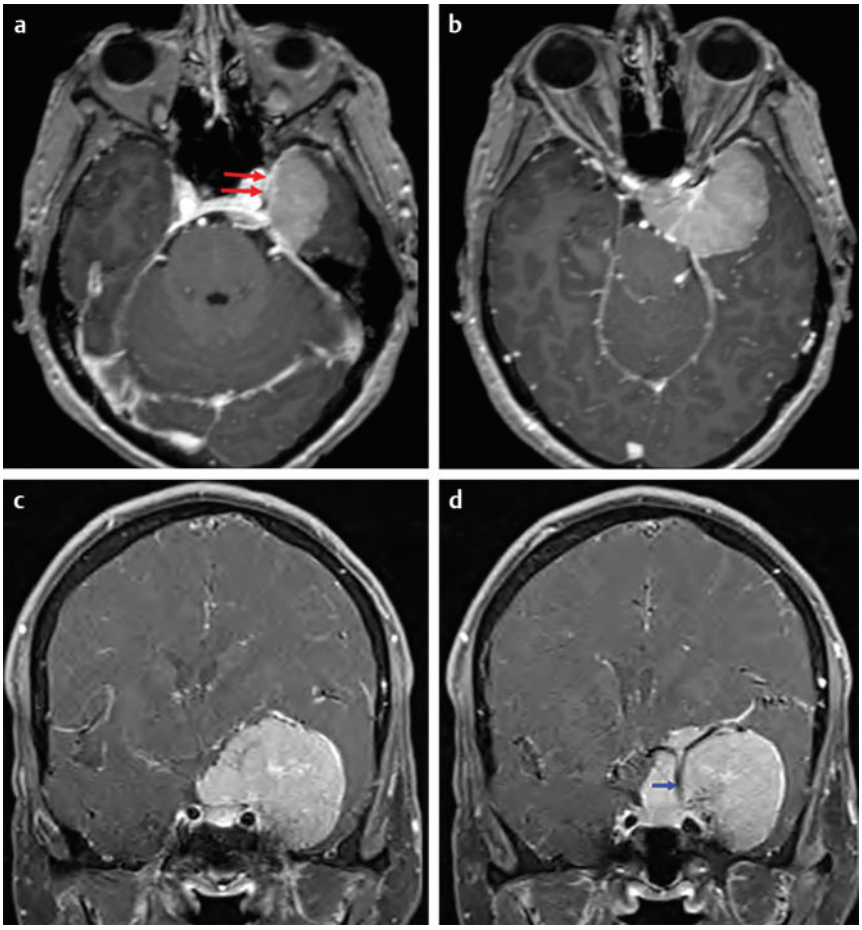


Fig. 6.15 Images of Case 3. (a–c) Axial and coronal MRI brain with contrast showing a large 5 × 4.6 × 4.8 cm meningioma that is centered on the left anterior clinoid process, sphenoid wing, and lateral wall of the cavernous sinus (*red arrows*). **(d)** The mass encased the left internal carotid (*blue arrow*) and middle cerebral arteries. The mass was also indenting the left side of the mesencephalon as well as compressing the left optic nerve and tract. The majority of this tumor was extracavernous and was, therefore, amenable to surgery.

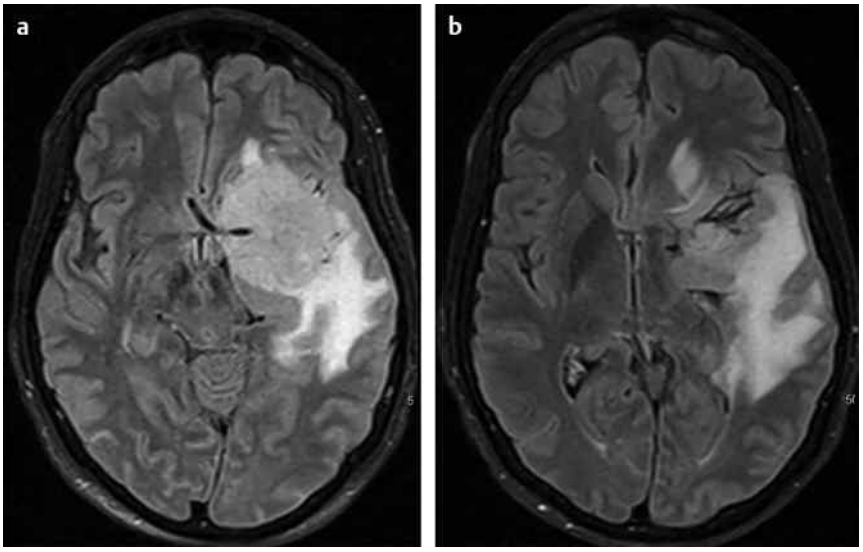


Fig. 6.16 **FLAIR images of Case 3. (a, b)** The peritumoral edema in the temporal lobe.

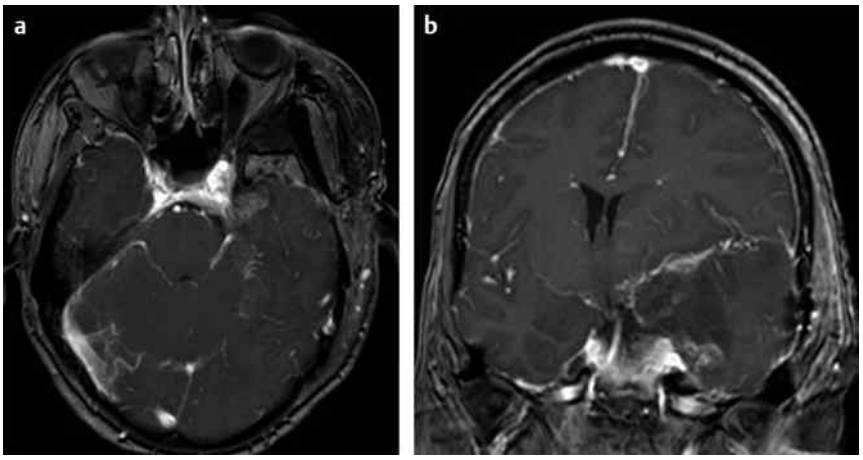


Fig. 6.17 **Postoperative images of Case 3. (a)** Axial and **(b)** coronal MRI images with contrast showed a total resection of the extracavernous meningioma using an FTOZ craniotomy.

peritumoral edema would be expected to be far more significant. Only a slow-growing tumor, such as a meningioma, would be found incidentally at this size.

This meningioma, while also in close proximity to the cavernous sinus, was anatomically very different from the one in Case 2. This tumor was attached to and infiltrated the lateral wall of the cavernous sinus and extended minimally into the lateral compartment. The majority of the tumor was in fact extracavernous. Unlike a holocavernous meningioma in Case 2, this tumor warranted a surgical intervention. It caused significant peritumoral edema (**Fig. 6.16**), and extended infratentorially to the prepontine cistern, compressing the midbrain and pons. The surgical goal was to relieve the mass effect on the brain, with preservation of the cranial nerves within the cavernous sinus. For the protection of the patient's intact neurological function, we did not intend to resect any intracavernous tumor confined within the lateral wall of the cavernous sinus.

For sphenocavernous meningiomas as in this patient's case, we tend to use some variant of a frontotemporal orbitozygomatic (FTOZ) craniotomy much like the authors of the previous section (**Fig. 6.17**). We believe that it decreases the amount of frontal and temporal lobe retraction required for exposure, especially for larger tumors. It should be stated of course that this is not the way we were taught to do this operation by Vinko Dolenc, who used a pterional craniotomy with a mid- and posterior orbitotomy and anterior clinoidectomy without FTOZ. It is also worth noting that similar to all skull base approach "add-ons," each has its own potential complications. For FTOZ this includes orbital pain, enophthalmos, pulsating exophthalmos, and the rare case of ocular injury, so it should be used judiciously.

■ Conclusion

The narrative description of mobilizing the lateral wall of the cavernous sinus combining the Kawase, the Hakuba, and the Dolenc methods is perhaps the most important concept for skull base/cavernous sinus surgeons to comprehend, and one which the senior author learned while operating with the three master surgeons. Start with this premise: "there really is no lateral wall of cavernous sinus." What there is instead is the temporal lobe dura, a continuation of the dura of the middle fossa covering the veins, nerve, and artery of the parasellar area and continuing on as the dura of the clinoids, the diaphragma sellae, the tuberculum, and even the clivus. With Hakuba, he always began the mobilization of the "lateral wall" at the junction of the superior orbital fissure with the anterior temporal dura and literally "pushed" it from front to back. There was no cutting necessary, and no dural opening. With Kawase, he would

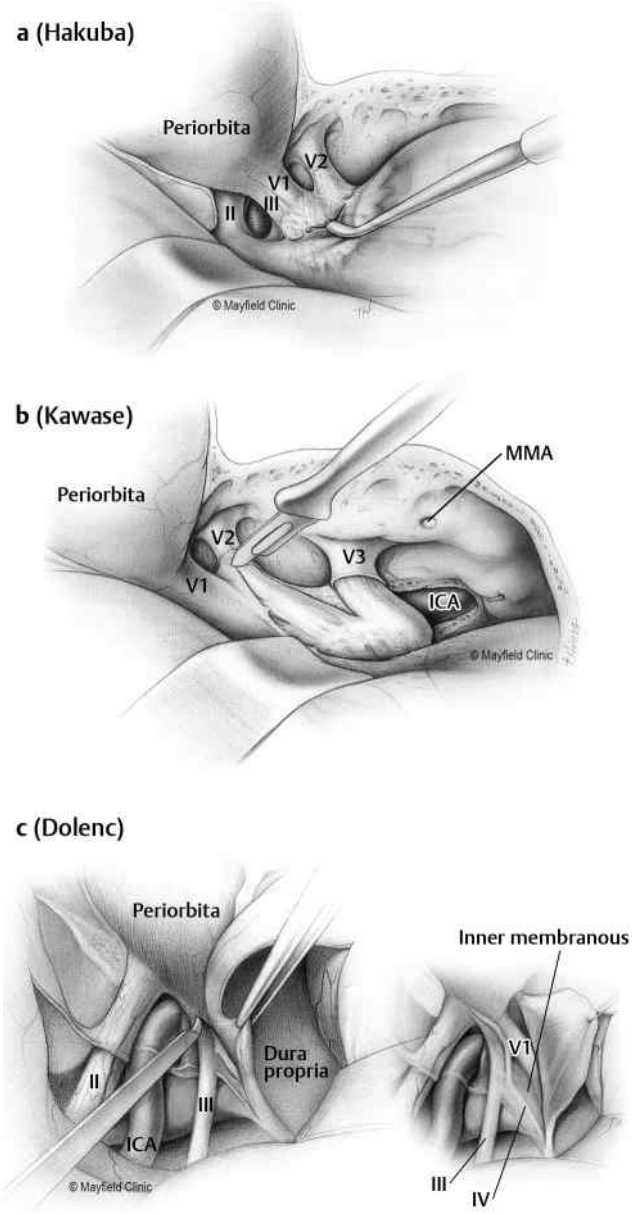


Fig. 6.18 Three methods used to mobilize the lateral wall of the cavernous sinus (AKA. medial temporal lobe dura). **(a)** The Hakuba method, **(b)** the Kawase method, **(c)** the Dolenc method. (Reproduced with permission from Mayfield clinic.)

release the dura sharply along the foramen ovale, bluntly along the foramen rotundum, and then dissect it bluntly up from bottom to top, first off Meckel's cave and then off the cavernous sinus. And with Dolenc, he would open the dura first and then find the oculomotor foramen behind the cavernous sinus. Then, he would cut all along the oculomotor nerve from back to front so the "lateral wall" could be peeled down from top to bottom (**Fig. 6.18a–c**).

With a sphenocavernous meningioma extending through the wall of the cavernous sinus or at least solidly invading it into the lateral compartment, all of these come together to peel the "lateral wall" off the cavernous sinus. In what order these maneuvers are made does not quite matter. Usually the configuration and texture of the tumor dictate which one works better and which is more challenging, as the surgeon explores each. In the end, the extracavernous tumor is resected, the cavernous sinus compartment is open and the compartment syndrome relieved, and the truly cavernous component of the tumor remains, for better or for worse.

The final point to be made, not only for cavernous sinus meningioma surgery but brain tumor surgery in general, is never forget to thank the tumor for its contributions to the outcome of the case. So often we are prone to blame the tumor for a bad outcome, or if not the tumor, the patients or their "protoplasm." Rarely do we acknowledge to our colleagues, our audience, or ourselves from the podium, that in some of our best cases, the softness of the tumor, its lack of adherence, or its configuration contributed to our great outcome, not necessarily in lieu of surgical skill but certainly in concert with it.

Recommended Reading

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Part III

Tumors of the Lateral Skull Base

7 Meckel's Cave *91*

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7 Meckel's Cave

R. Tushar Jha, H. Jeffrey Kim, and Walter C. Jean

Keywords: anterior petrosectomy, gasserian ganglion, endoscopic transpterygoid approach, vidian nerve, paraclival carotid artery

■ Case Presentation

A 45-year-old woman with a past medical history of hypertension presented to the emergency department with new onset of left-sided jaw and facial numbness, as well as intermittent mild left frontal headache. On physical examination, she was noted to have dysesthesias over the left side of her face and jaw, and slight tongue deviation to the right. Magnetic resonance imaging (MRI) of the brain revealed an extra-axial, dumbbell-shaped, left middle fossa lesion. The mass was considered "benign" but compressed the pons. The patient was told to obtain serial brain MRI scans at 3-month, 6-month, and 1-year intervals. These MRIs revealed interval growth of the lesion, and the patient was referred to our skull base division.

Questions

1. What is the differential diagnosis of the MRI finding?
2. In retrospect, was the period of observation a rational option?
3. How does the unique shape of the tumor influence the selection of surgical approach?

■ Diagnosis and Assessment

At the time of presentation to our skull base clinic, the MRI showed a well-demarcated, heterogeneously enhancing, dumbbell-shaped mass in the Meckel cave region of the left middle cranial fossa (**Fig. 7.1**). The differential diagnosis was between a meningioma and trigeminal schwannoma, but the latter was favored as it is the most common neoplastic lesion of Meckel's cave. These lesions make up about 33% of Meckel's cave tumors and are classified into three types based on their pattern of growth. Type I lesions are confined to the middle fossa. Type II lesions are dumbbell-shaped due to anterior and posterior extension into the cavernous sinus and posterior fossa, respectively. Type III lesions grow primarily posteriorly through the porus trigeminus into the posterior fossa and exhibit minimal extension into the middle fossa. Other neoplastic lesions are far less common, but still worthy of consideration. These include retrograde extension of head and neck tumors, epidermoid cyst, lipoma, and petrous apex cephalocele.

The dumbbell-shaped Meckel's cave mass in this case measured 3.5 × 3.2 × 2.4 cm in anterior–posterior, transverse, and height. There was extension into the left prepontine cistern with moderate mass effect on the pons. The pontine compression had increased when compared to the patient's initial MRI 1 year previously. The mass extended anteriorly along the middle cranial fossa next to the left cavernous sinus, posteriorly through the porus trigeminus into the prepontine cistern, and inferiorly into the left upper cerebellopontine angle (CPA). In the CPA, the tumor was abutting the left seventh and eight cranial nerve complex. These findings favored the radiographic diagnosis of a type II trigeminal schwannoma.

Trigeminal schwannomas are benign tumors that typically exhibit slow growth. Given the indolent nature of these lesions and the complexity of their surgical management, a period of observation is not unreasonable for most patients at the time of diagnosis. However, the brainstem compression at diagnosis should have raised concern for our patient. When she arrived at our clinic, with radiographic progression over a 1-year period and worsening brainstem compression, few would argue that surgery was the only viable option at this point.

■ Anatomical and Therapeutic Considerations

Prior to considering the various surgical approaches to this tumor, it is important to take into account the anatomy of the middle fossa, Meckel's cave, cavernous sinus, and the

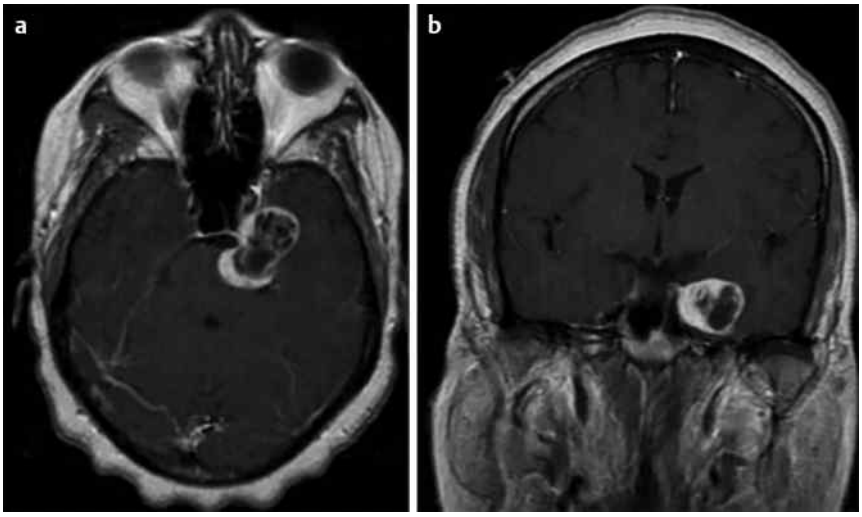


Fig. 7.1 Preoperative MRI. (a) Axial and **(b)** coronal MRI with gadolinium showing a dumbbell-shaped enhancing mass in the region of the left Meckel's cave.

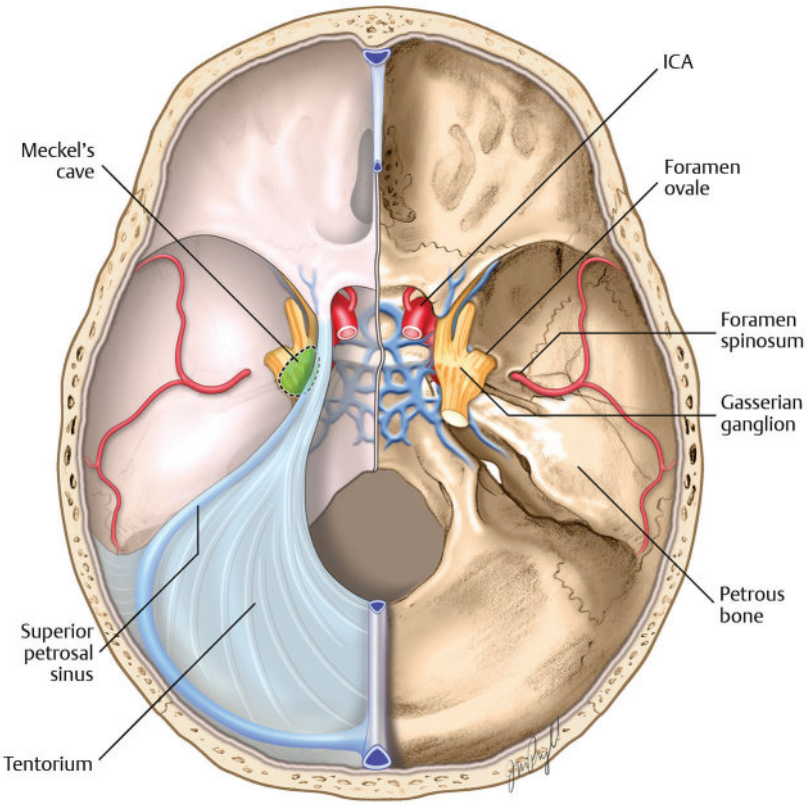


Fig. 7.2 Boundaries of the middle fossa. Schematic illustration showing the boundaries of the middle fossa and relationship of Meckel's cave, petrous bone, superior petrosal sinus, and cavernous carotid artery to each other.

relationships of these structures to one another (**Fig. 7.2**). The middle fossa is bound anteriorly by both wings of the sphenoid bone, posteriorly by the petrous temporal bone, laterally by the squamous part of the temporal bone, and medially by the sella turcica. Just lateral to the sella turcica is the cavernous sinus. The detailed anatomy of the cavernous sinus is beyond the scope of the current discussion but relevant to our patient's tumor; it is important to highlight the two-layer structure of the lateral wall. The superficial, thicker, layer is contiguous with the dura propria of the temporal lobe and forms the dorsal wall of Meckel's cave. The deeper reticular layer is contiguous with the epineurium of cranial nerves III, IV, and VI. The separation of the two layers will be important in surgery.

Meckel's cave sits posterior and lateral to the cavernous sinus (**Fig. 7.3**). It is a three-fingered glove-shaped dural recess containing the gasserian ganglion, V1, V2, and V3 sensory divisions, and the motor rootlets of the trigeminal nerve. Its opening or "mouth" is situated at the petrous apex just below the superior petrosal sinus (SPS). Meckel's cave then extends anteriorly and inferiorly down the anterior face of the petrous bone until the sensory divisions and motor rootlets reach their respective foramina. The petrous and lacerum segments (C2, C3) of the internal carotid artery (ICA) courses within the petrous bone under Meckel's cave.

Our patient's tumor expanded Meckel's cave. In the middle fossa, the anterior "lobe" of the dumbbell tumor involved the lateral wall of the cavernous sinus. The posterior "lobe" of the dumbbell was in the posterior fossa occupying the prepontine cistern. Inferiorly, this "lobe" abutted the VII/VIII nerve complex at the internal auditory canal (IAC).

The most threatening feature of the patient's tumor was its mass effect on the brainstem. Therefore, the primary surgical goal must be to decompress the brainstem. The secondary goal was

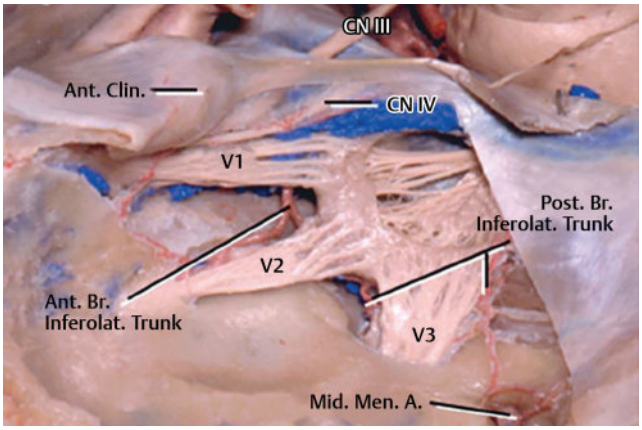


Fig. 7.3 Neurovascular relationship of the middle fossa floor and Meckel's cave (left side). Tentorium and dura of Meckel's cave have been removed. A., artery; Ant., anterior; Br., branch; Clin., clinoid; CN, cranial nerve; Inferolat., inferolateral; Men., meningeal; Mid., middle; Post., posterior. (Reproduced from Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery. 1st ed. Thieme; 2011.)

total resection of the lesion. If the histopathological identity of the tumor was in fact a grade I meningioma or schwannoma, then a total resection would potentially be curative for our patient. Intraoperatively, if portions of the tumor proved to be adherent to critical neurovascular structures, then leaving a small residual would be acceptable as long as the primary goal was met.

Options of Approach

Meckel's cave is unique in skull base surgery, as it can be reasonably accessed with several surgical corridors. In broad

terms, the main corridors are anterior, lateral, and posterior, and each has its strengths and weaknesses.

The anterior approach is performed through an endoscopic endonasal transpterygoid approach. The technique requires complete ethmoidectomy, sphenoidotomy, and removal of the posterior wall of the maxillary sinus to gain access into the pterygopalatine fossa (PPF). Then, by following the vidian nerve to identify the pterygoid plate, further bony drilling in this area (with or without sacrificing the vidian nerve and artery) allows enough exposure to follow V2 through foramen rotundum into Meckel's cave (**Fig. 7.4**).

With this technique, there is no need for temporal lobe retraction, which is a frequent cause of neurological injury associated with the lateral approaches. However, the approach requires delicate drilling to expose the paraclival carotid artery and can be technically challenging. Specifically, for our patient, the biggest disadvantage of the endonasal route is the long reach and limited access into the posterior fossa. As the primary goal for her is brainstem decompression, the transpterygoid approach was not her best option.

If the anterior approach has difficulty accessing the brainstem, the posterior approach must logically be considered next as this option puts the surgeon closest to the brainstem. Through the standard retrosigmoid craniotomy, exposure of Meckel's cave is usually blocked by the suprameatal tubercle, a hump of bone at the superior lip of the internal acoustic meatus. However, by removing this tubercle with the drill, access can be gained not only to Meckel's cave, but also to the petrous apex and posterior parts of the middle fossa.

The strength of this retrosigmoid intradural suprameatal approach is the early exposure of the trigeminal nerve at the brainstem, allowing for efficient brainstem decompression. However, for our patient, the attachment of the tumor to the lateral wall of the cavernous sinus would pose significant problems for the posterior approach, even with endoscopic-assisted access into the middle fossa. Just as the anterior approach would have difficulty reaching the posterior lobe of the dumbbell, the posterior approach would have the same problem with the anterior lobe.

Our considerations turn next to the lateral trajectory. This route is centered on a middle fossa craniotomy and subtemporal extradural approach, which would theoretically provide equal access to both lobes of a dumbbell-shaped tumor, balanced at the anterior petrous apex. A middle fossa craniotomy would also gain access to the lateral wall of the cavernous sinus, allowing the mobilization of the two layers of this wall away from each other to detach the tumor from the cavernous sinus. However, what about the posterior fossa? By definition, a middle fossa craniotomy is supratentorial, so what about the exposure into the posterior fossa to accomplish the primary goal of decompressing the brainstem?

The answer to these questions lies with the addition of an anterior petrosectomy. The boundaries of Kawase's quadrilateral are the greater superficial petrosal nerve (GSPN) laterally, mandibular division of cranial nerve V anteriorly, the SPS and petrous ridge medially, and the internal acoustic canal (IAC) posteroinferiorly (**Fig. 7.5**). By supplementing the middle fossa craniotomy with drilling at the Kawase quadrilateral, the posterior and inferior parts of our patient's tumor can be exposed. Furthermore, with the anterior petrosectomy, the surgeon gains control of the SPS and the lateral edge of the tentorium cerebelli. If the latter is cut, further opening into posterior fossa can be gained.

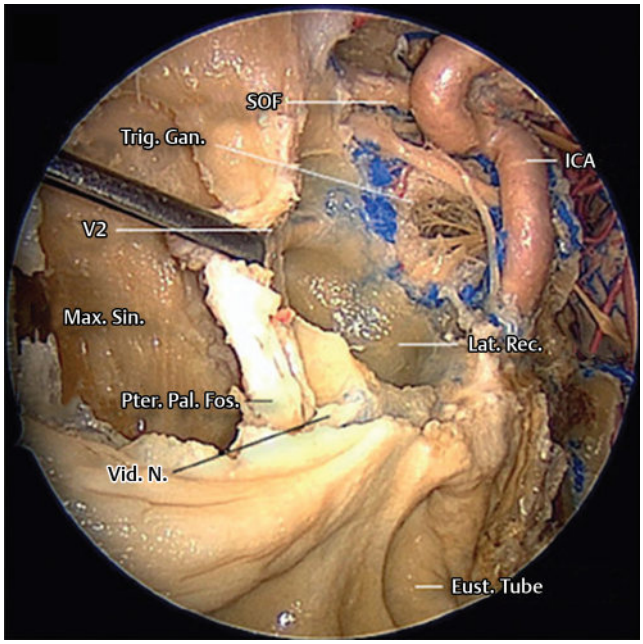
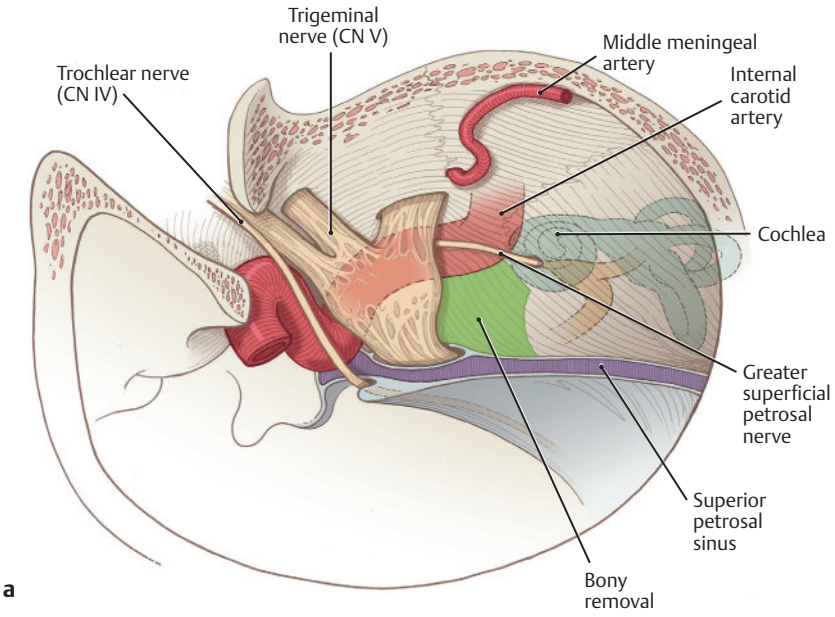


Fig. 7.4 Overview of the endoscopic approach to Meckel's cave. Removal of the posterior wall of the maxillary sinus exposes the pterygopalatine fossa, where the vidian canal and foramen rotundum can be identified. The pterygoid process has to be drilled out to access the lateral recess of the sphenoid sinus. The vidian nerve runs in the floor of the lateral recess, and the maxillary nerve (V2) courses in the lateral wall. Both nerves tend to converge as they travel posteriorly, and are used as surgical landmarks that guide the approach. The vidian nerve, which can be preserved in selected cases, is followed posteriorly until it meets the lateral aspect of the internal carotid artery at the foramen lacerum, and the maxillary nerve is followed posteriorly where it joins the trigeminal ganglion, located posterolateral to the paraclival segment of the internal carotid artery. The superior orbital fissure, which is the anterior continuation of the cavernous sinus, is located anterosuperior to Meckel's cave and anterolateral to the parasellar segment of the internal carotid artery. Eust., Eustachian; Fos., fossa; Gan., ganglion; ICA, internal carotid artery; Lat., lateral; Max., maxillary; N., nerve; Pal., palatine; Pter., pterygoid; Rec., recess; Sin., sinus; SOF, superior orbital fissure; Trig., trigeminal; Vid., vidian. (Reproduced from Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery. 1st ed. Thieme; 2011.)

The main disadvantage of the lateral route is that forceful retraction of the temporal lobe is needed to fully expose the petrous ridge. Furthermore, drilling during the anterior petrosectomy puts the cochlear and petrous carotid at risk, not to mention the cranial nerve VII/VIII complex at the posteroinferior limits of the drilling.

Approach of Choice

Considering all the factors mentioned above, we had a difficult decision to make for our patient. The anterior endonasal approach was deemed too risky for the contents of the posterior fossa, and again, given that the primary goal is decompression of her brainstem, the endoscopic endonasal approach (EEA) was ruled out. The posterior retrosigmoid approach required too long a reach to access the middle fossa, so the final decision was made to use the lateral trajectory. A middle fossa craniotomy will be supplemented with an anterior petrosectomy with the



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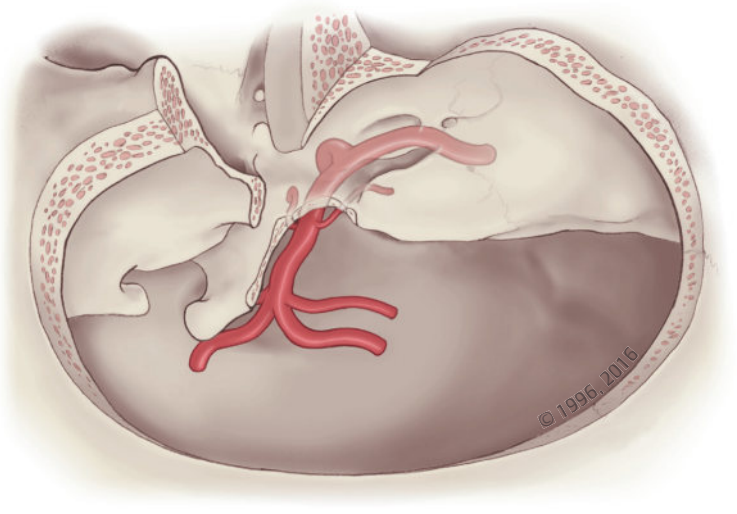

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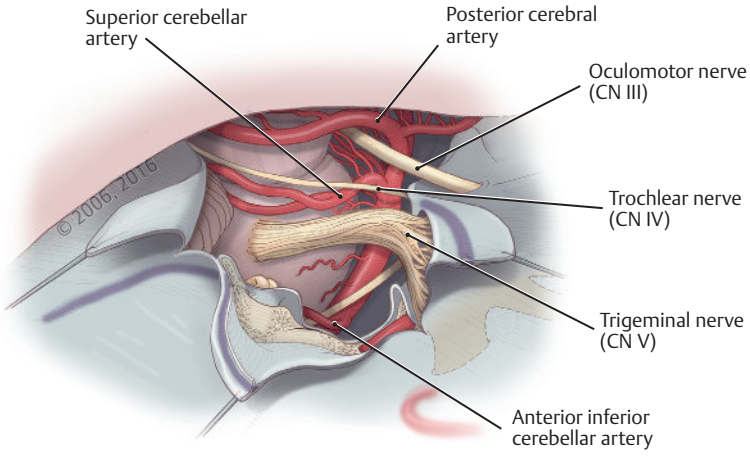

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Fig. 7.5 Anterior petrosectomy. (a) Anatomical landmarks for drilling of the Kawase approach. The anterior margin for drilling is the trigeminal nerve, the lateral margin is the greater superficial petrosal nerve, the inferior margin is the internal carotid artery, and the medial margin is the superior petrosal sinus. Posteriorly, the cochlea should be preserved to prevent injury to hearing. The extent of bony removal is depicted (*green area*). **(b)** The anterior petrosectomy provides enhanced exposure to the ventrolateral brainstem. Drilling of the medial apex of the petrous bone increases exposure to the clivus and the upper basilar artery. **(c)** Final anatomical exposure afforded by the anterior petrosectomy. (Reproduced from Spetzler R, et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

aim of decompressing the brainstem, and mobilization of the lateral wall of the cavernous sinus to potentially fully resect this patient's tumor to prevent recurrence.

Questions

1. How are the boundaries of Kawase's quadrilateral identified during surgery?
2. What are the critical structures that may be "at risk" during an anterior petrosectomy?
3. If incision of the tentorium is necessary, how and where does one start? What structures are "at risk" during this incision?

■ Description of the Technique

Prior to positioning the patient for the surgical approach, the patient was intubated under general anesthesia and placed in the lateral position for placement of a lumbar drain. This will be used during surgery for brain relaxation. The patient was then positioned supine with a shoulder roll under the left shoulder. The thorax was elevated about 10 degrees to facilitate venous drainage. The head was rotated toward the right shoulder to a position almost parallel to the floor. The neck was slightly extended and the head was inferiorly tilted so that the zygoma was the highest point in the surgical field. This head position allowed gravity to assist the mobilization of the temporal lobe away from the middle fossa floor. Facial nerve monitoring electrodes were placed prior to sterile draping (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: subtemporal

Craniotomy: middle fossa

Modifiers: anterior petrosectomy

A question mark–shaped incision was then made starting from the tragus, curving upward, posteriorly, and anteriorly again.

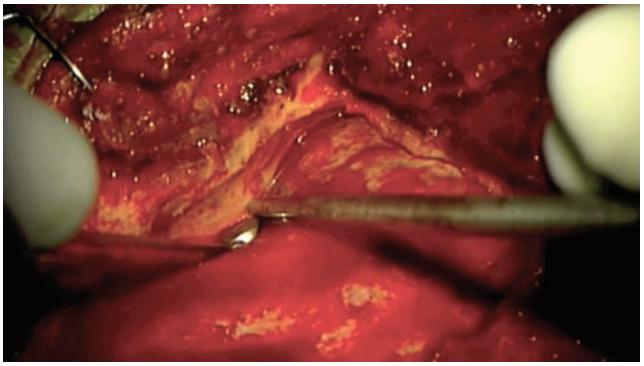


Fig. 7.6 Middle fossa floor. Extradural elevation of the temporal lobe dura off the middle fossa floor was performed with the Penfield number 1 dissector.

The temporalis muscle was incised and reflected with the skin flap anteriorly inferiorly. A posterior myofascial cuff was left to assist with temporalis closure at the end of the surgery. The temporal root of the zygoma was exposed and a burr hole was placed just above it. A 5×5 cm square-shaped middle fossa craniotomy was then made, and the remaining lip of bone at the inferior aspect of the craniotomy was rongeured down flush to the middle cranial fossa floor. At this point, the lumbar drain was opened to drain cerebrospinal fluid (CSF) to facilitate brain relaxation.

The Anterior Petrosectomy

The temporal lobe was elevated in an extradural fashion using a Penfield number 1 dissector (**Fig. 7.6**) until the superior bony petrous ridge was delineated. The middle meningeal artery, found at the foramen spinosum, was coagulated and then divided, allowing further elevation of the temporal lobe dura. The GSPN and V3 were then visualized, and the identity of the former was confirmed using the facial nerve stimulator. The anterior petrosectomy started with drilling posterior to V3 and medial to the GSPN, and it became evident that the tumor had eroded much of Kawase's quadrilateral (**Fig. 7.7**).

The drilling uncovered the ICA in its horizontal petrous portion inferolateral to the GSPN. It was noted that the GSPN and the arcuate eminence (AE) formed the expected 120-degree angle. The direction of the IAC was estimated by bisecting this angle, and the drilling proceeded over the meatal plane to identify the entire course of the IAC. The drilling continued posteriorly and inferiorly toward the inferior petrosal sinus by following the dura of Meckel's cave into the posterior fossa. The facial nerve (cranial nerve VII) was then uncovered as it traveled from the fundal area of the IAC (lateral) to the GSPN and the geniculate ganglion, while avoiding the cochlea situated just anterior to the IAC fundus. The cortical bone at the junction of the ICA and IAC was carefully removed until the lighter color and harder bone of the otic capsule were encountered. The cochlea

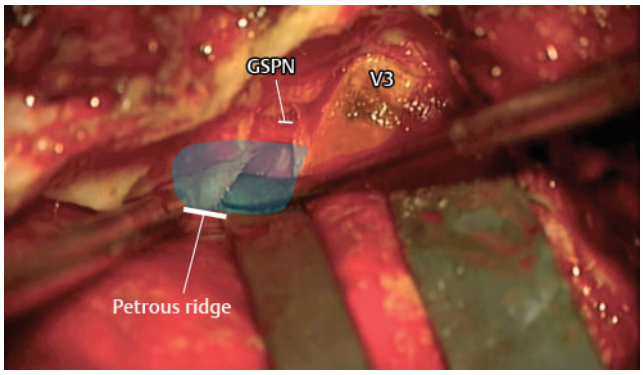


Fig. 7.7 Kawase's quadrilateral. View through the operating microscope of the middle fossa floor prior to anterior petrosectomy. The faintly yellow shaded area is V3 and the blue shaded area denotes Kawase's quadrilateral. Note that the dissector is showing that the petrous ridge that was partially eroded by the patient's tumor. GSPN, greater superficial petrosal nerve.

was "blue-lined" and after this the anterior petrosectomy was complete (see **Operative Setup**).

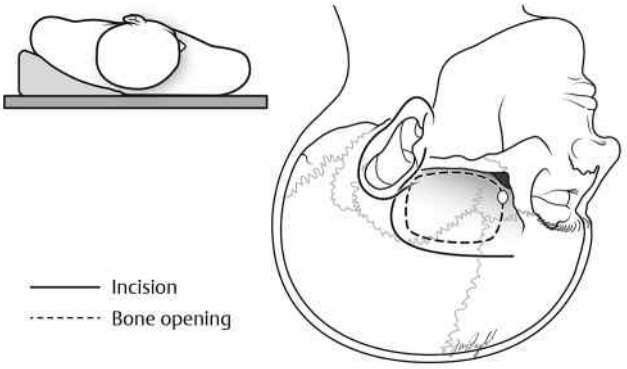
Operative Setup

Position: supine with head turned almost 90 degrees to the right

Incision: question mark, starting just anterior to tragus

Bone Opening: middle fossa, approximately 5 cm^2

Durotomy: parallel to the middle fossa floor, parallel to brainstem



The Posterior Fossa

The dura was opened next to the middle fossa floor and parallel to it. Then, by elevating the temporal lobe, the tentorium was exposed. The posterior fossa dura, exposed via the anterior petrosectomy, was opened parallel to the brainstem. Where the two perpendicular dural incisions met, the SPS was coagulated and divided.

The tumor was then debulked using an ultrasonic aspirator. Cranial nerve V was identified at its brainstem origin and was found to be completely entangled with the tumor. As such, this was detached from the brainstem by incising the involved rootlet. The tentorium, already cut where the SPS was divided, was further incised medially until the incisura, exposing the trochlear nerve (**Fig. 7.8**). The final tentorial cut released the tumor significantly, allowing the tumor to be dissected off the pons (**Fig. 7.9**). The tumor was completely removed from the posterior fossa at this point and brainstem decompression was achieved.

The Middle Fossa

The anterior half of the tumor was debulked using the ultrasonic aspirator to gain further exposure toward the cavernous sinus. The last remnants of the tumor within Meckel's cave and adjacent to the cavernous sinus were completely removed by mobilizing the lateral wall of the cavernous sinus. To achieve this, V2 and V3 were identified at foramen rotundum and ovale, respectively, and their outer dural covering—the dura propria—was dissected away while maintaining the integrity of the inner reticular layer. In such a way, the tumor adherent to the cavernous sinus wall was removed with minimal harm to the cranial nerves of the cavernous sinus, and safeguarding the intracavernous ICA.

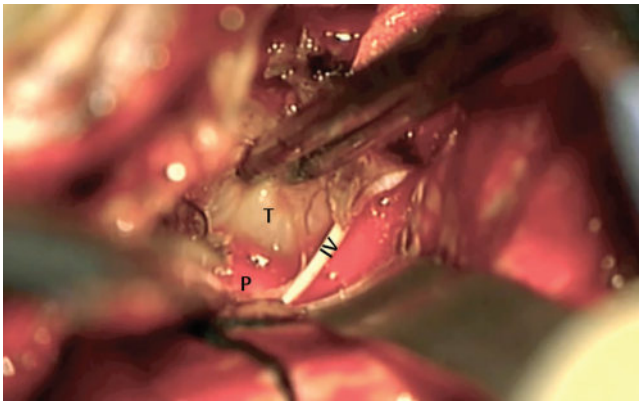


Fig. 7.9 Brainstem decompression. With the incision of the tentorium complete, the tumor (T) can finally be dissected away from the pons (P). IV, trochlear nerve.

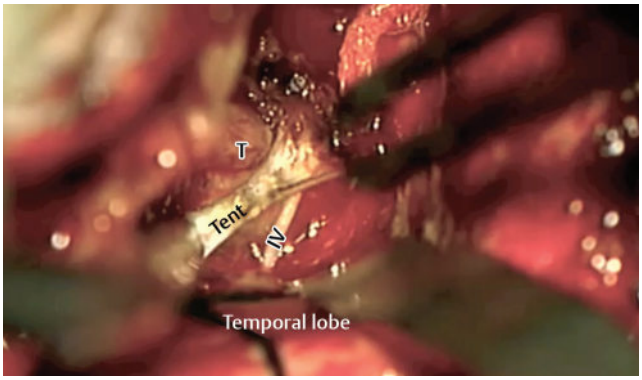


Fig. 7.8 Tentorial incision. With the incision of the tentorium nearing completion, one can see the trochlear nerve (IV) at the incisura. The release of the tentorium reduces the compressive forces of the tumor (T) against the brainstem.

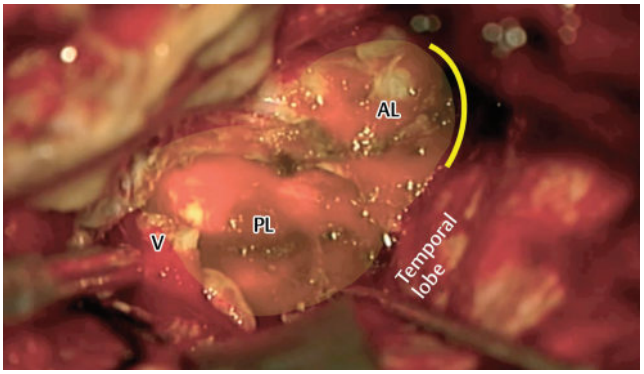


Fig. 7.10 End of resection. The faintly yellow area denotes tumor "bed" after the complete removal of the mass. The *yellow line* indicates the lateral wall of the cavernous sinus from which the tumor was detached. AL, anterior lobe; PL, posterior lobe of the dumbbell tumor; V, stump of trigeminal nerve.

At the end of the resection process, all visible tumor had been removed (**Fig. 7.10**). The temporal lobe was unharmed. The facial nerve responded with 0.5 mA of stimulation from the brainstem underneath the dural covering. The large air cells encountered during the initial approach were packed with collagen-based dural substitute, and a piece of temporalis muscle was harvested and placed over the petrous apex defect. The dural edges were approximated together with sutures and exposed mastoid air cells were plugged with wax. After the bone flap was replaced and skin closed, the lumbar drain was left in place during the immediate postoperative period.

An edited version of the video from this operation accompanies this text (**Video 7.1**).

Surgical Pearls

1. An anterior petrosectomy is a very "deep" undertaking, as there is a significant distance between the craniotomy opening and where the drilling takes place. As such, there is also a lot of temporal dura that must be elevated, and protection of the temporal lobe is paramount. Positioning the head to allow gravity to assist the retraction mitigates the risk of injury. The use of a lumbar drain is controversial, but it is the senior authors' custom to use this to lessen the necessary retraction force.
2. Cochlear injury is unfortunately common during anterior petrosectomy, and it is the basal turn of the cochlea that is most commonly violated during this approach. Important landmarks for identifying the cochlea include GSPN, geniculate ganglion, IAC, and the petrous ICA; however, no definite landmark for the cochlea is established because extensive anatomical variations exist between patients. The basal turn is typically medial to the geniculate ganglion in the angle between the labyrinthine segment of the facial nerve and the GSPN (**Fig. 7.11**). The mean distance between the medial edge of the basal turn and the medial side of the labyrinthine segment of the facial nerve is 4.4 mm (range 3.7–5.1 mm). During anterior petrosectomy, the bone can be removed laterally until the hard otic capsule is identified. The otic capsule appears distinctly firmer and lighter in color than the typical petrous apex bone. After the GSPN and labyrinthine segment of the facial nerve is delineated toward the fundus of the IAC, *extreme caution* should be taken to avoid injury to the basal turn of the cochlea when drilling deep and medially to the labyrinth segment of the facial nerve and posterior to the posterior genu of the petrous ICA.
3. Fully incising the tentorium is critical to this operation, as without it, the tentorium compresses the tumor against the brainstem. The tentorium incision starts at the SPS. By opening the posterior and middle fossa dura perpendicular to each other, one never needs to search for the SPS as it is inevitably where the two cuts meet. Once the SPS is coagulated, the location to cut the tentorium becomes "self-evident."

■ Aftercare

The patient did well after her surgery. Her head of the bed was elevated to 30 degrees and lumbar drain was opened to drain CSF at a continuous rate of 5 mL/ho to minimize the chance of CSF leakage. Her initial postoperative examination revealed left facial weakness (House–Brackmann II/VI), and, as expected, left facial numbness. Dexamethasone was started to help manage the patient's facial weakness. Deep vein thrombosis prophylaxis was instituted the afternoon following surgery.

The patient's lumbar drain was clamped on day 5 after surgery and was removed on day 6 as she had no evidence of a CSF leak. The patient was discharged home on day 7 after surgery and by then her facial weakness had completely resolved. Steroids were stopped. MRI after her surgery showed a total resection with decompression of the brainstem (**Fig. 7.12**). The histopathological diagnosis from the resection was a schwannoma with an MIB index (Ki-67) of 1 to 3%.

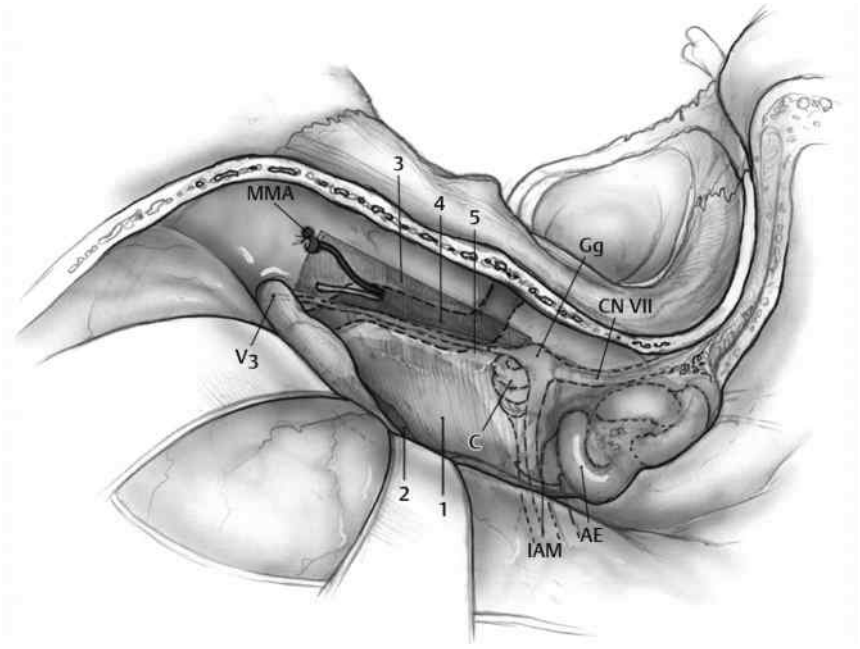


Fig. 7.11 Cochlea at the middle fossa floor. Note the location of the cochlea. The basal turn is medial to the geniculate ganglion, in the angle of the labyrinthine segment of the facial nerve and the GSPN. 1, Kawase's triangle; 2, trigeminal impression; 3, Glasscock's triangle; 4, petrous carotid; 5, GSPN; AE, arcuate eminence overlying the anterior semicircular canal; C, cochlea; Gg, geniculate ganglion; IAM, internal auditory meatus; MMA, middle meningeal artery; V3, mandibular nerve. (Reproduced from Nader R et al, eds. Neurosurgery Tricks of the Trade. Thieme; 2014.)

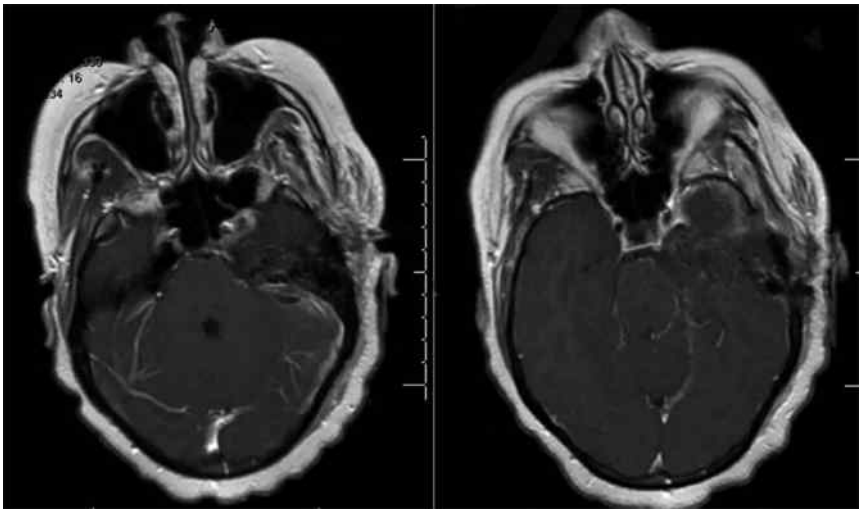


Fig. 7.12 Postoperative MRI. Axial MRI with gadolinium showing a complete resection of the Meckel's cave schwannoma with decompression of the brainstem.

■ Possible Complications and Associated Management

Complications from this operation can be separated into those related to the approach or the resection. An anterior petrosectomy can be complicated by postoperative CSF leak, cranial nerve VII injury, hearing loss, and temporal lobe contusion. CSF leaks and failure in wound healing can be limited by appropriately plugging any exposed air cells with wax, grafted tissue, or dural substitute. Injury to the cranial nerve VII/VIII complex can be minimized by accurately delineating the GSPN, AE, and then the IAC that bisects the angle formed by the former two structures. Drilling away the bone overlying the IAC with a diamond burr under copious irrigation can minimize infiltrating the dural sheath of the IAC and thermal injury to the nerves. If facial nerve palsy happens despite these precautions, it can be managed with a short postoperative course of steroids. To minimize the risk of hearing loss, the cochlea must be avoided during the drilling of the petrous apex. This has been discussed in the "Surgical Pearls" section. The temporal lobe can be contused if overvigorous retraction is used to expose the floor of the middle fossa. CSF diversion and using the least retraction necessary are potential ways to avoid this.

Resection-related complication can be minimized by meticulous microsurgical techniques while maneuvering around the brainstem. Cranial nerves IV through VIII can be adherent to the tumor capsule in varying degrees in the posterior fossa and tentorial incisura, and if so, must be carefully dissected free. Mobilization of the lateral wall of the cavernous sinus can not only maximize the degree of tumor removal, but also moves the cavernous sinus out of harm's way. "Wandering" into the cavernous sinus with the resection tool can have catastrophic consequences especially when the ICA is penetrated.

Perspective

Alexandre B. Todeschini, Bradley A. Otto, Ricardo L. Carrau, and Daniel M. Prevedello

■ Introduction

Tumors involving Meckel's cave are complex surgical challenges because of the delicate structures surrounding the area. No single approach provides unlimited access to the entire region, and lesions in the Meckel's cave are frequently multicompartmental, adding to the complexity. As always, clearly established surgical goals and a thorough knowledge of anatomy and techniques are needed to select the ideal access through the safest corridor for each lesion, and, often, a combination of two or more approaches is necessary to achieve a satisfactory outcome.

For the case presented in the preceding section, the objectives for the surgical approach were well-defined as was the anatomy of the case. The middle fossa approach was the best choice for that lesion, as it allowed the access to the anterior portion of the tumor directly and extradurally through a middle fossa peeling. The posterior aspect can be followed through the tumor itself or through an anterior petrosectomy (Shiobara–Kawase approach).

We will present an alternative case and focus on the EEA.

■ Case Presentation

A 67-year-old man presented to our clinic with numbness on the right side of his face (V1–V3) and mild headaches. Imaging studies showed a large heterogeneously enhancing mass, centered in the region of Meckel's cave and medial aspect of the right middle cranial fossa, with extensions to adjacent areas including to the posterior fossa. The most likely diagnosis was a trigeminal schwannoma (**Fig. 7.13**).

■ Anatomical and Therapeutic Considerations

Comparing the images of this case with the previous one, we note that the first case had a larger posterior fossa component and that the lesion was behind the ICA without protrusion into the sphenoid sinus. In contrast, our case had only a small component in the posterior fossa, but did project into the lateral recess of the sphenoid sinus anterior to the ICA, making the EEA the ideal choice for it (**Fig. 7.13**).

Options for Approach

When the main component of the trigeminal schwannoma is in the middle fossa, but guarded by the ICA anteriorly, a middle fossa approach with extradural peeling is the approach of choice. This option has been thoroughly discussed. When the main component of the trigeminal schwannoma is in the posterior fossa with only a minor portion in Meckel's cave, then a retrosigmoid approach can be used. In this situation, Meckel's cave itself can be reached by drilling of the suprameatal tubercle, often performed with endoscopic assistance.

Another treatment strategy which is gaining popularity among skull base surgeons, especially for multicompartmental benign lesions, is a staged approach, whereby multiple surgical procedures are planned to remove the accessible portions of tumor. When the target portion is removed, the operation is halted, respecting the safety limits of the approach. A second stage, using a separate approach specifically chosen for the remaining portion of the tumor, is done after an adequate time

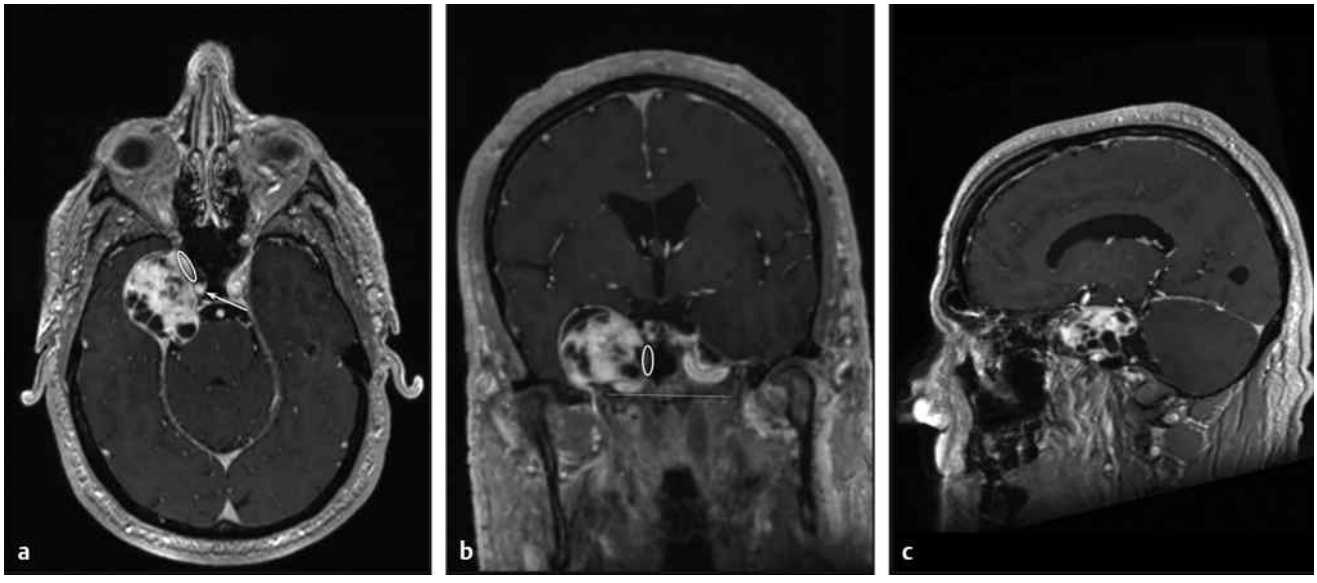


Fig. 7.13 Preoperative images. Contrast-enhanced MRIs. **(a)** Axial view. Note the internal carotid artery (ICA) was pushed posteriorly (*arrow*) creating an unobstructed trajectory to the tumor (*oval*). **(b)** Coronal view. A similar access to the tumor can be identified (*oval*), further aided and increased by the caudal displacement of the ICA when compared to the contralateral (*thin line*). **(c)** Sagittal view. The posterior fossa component was small.

interval from the first stage. The advantage of this strategy is in harnessing the strength of each approach, as only the portion of the tumor most accessible to it is removed. The weakness is subjecting the patient to multiple surgeries, possibly compounding the risks of each stage.

Approach of Choice

Returning to our patient, his tumor projected into the sphenoid sinus anterior to the carotid artery, and this anatomical nuance can be exploited for direct surgical access. Indeed, the EEA, using the anteromedial corridor to access Meckel's cave, is uniquely suitable for this tumor (**Fig. 7.14**).

This technique starts with an endoscopic transpterygoid approach, and the vidian canal is followed until the petrous ICA is located. The bone covering the paraclival and parasellar carotid is removed, followed by extensive removal of bone around foramen rotundum. Access to Meckel's cave is thusly achieved through the quadrangular space which is bounded by ICA inferiorly and medially, V2 laterally, and the abducens nerve superiorly. The resulting corridor provides a direct ventral trajectory to reach the anterior and inferomedial aspects of Meckel's cave. No other corridors can provide this access, because for lesions in this area, the trigeminal nerve itself blocks the approach from all other directions.

■ Description of the Technique

The approach was performed with an otolaryngologist and neurosurgeon working together using two-surgeon four-handed technique. The patient was positioned supine with the head tilted left and slightly turned to the right in a rigid three-pin head holder. Topical oxymetazoline was applied to the nasal cavity for vasoconstriction, and image guidance was utilized.

To begin, the transpterygoid approach started with resection of the right middle turbinate, lateralization of the left middle turbinate, and outfracture of the inferior turbinate bilaterally. A pedicled nasoseptal flap was elevated for later use in reconstruction, and this was done from the contralateral side of the approach to make sure the arterial pedicle would be preserved. A posterior nasal septectomy (1.5 cm), wide bilateral sphenoidotomies, and posterior ethmoidectomies were done. The basopharyngeal fascia was then stripped away from the undersurface of the sphenoid floor, which was drilled down to the level of the clival recess (**Fig. 7.15a**). Next, the vessels emerging from the sphenopalatine foramen (sphenopalatine and posterior nasal arteries) were isolated and coagulated (hence, the contralateral nasoseptal pedicle). A posterior maxillary antrostomy was then performed, and the medial pterygoid plate was identified after lateralization of the contents of the PPF while keeping the periosteum intact.

At this point, the vidian nerve and artery bundle can be clearly seen originating from the vidian canal along the base of the pterygoid plates (**Fig. 7.16**). By following the sphenoid floor from a medial-to-lateral direction, the vidian canal is usually found at the junction of the sphenoid floor as it disappears laterally and transitions to the medial pterygoid wedge (**Fig. 7.15b**). The most critical landmark for transpterygoid approaches is the petrous ICA, and the vidian canal is the guide to identify it. Once the canal was identified, drilling proceeded cautiously

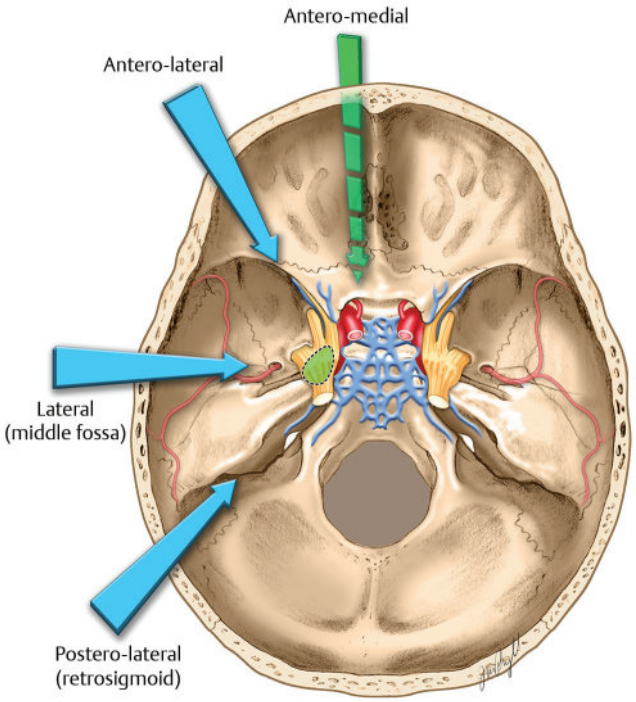


Fig. 7.14 Options for approach. The endoscopic endonasal transpterygoid approach using the anteromedial corridor is uniquely suited for Meckel's cave tumors with small posterior fossa components, projected into the sphenoid sinus, that lie anterior to the internal carotid artery. Tumors that predominantly occupy the inferomedial aspect of Meckel's cave are best approached through the endoscopic endonasal approach, as the trigeminal nerve blocks the approach from all other directions. Green area, Meckel's cave.

along its inferior and medial aspect in an anteroposterior direction toward the foramen lacerum. When the position of the anterior genu (petrous–paraclival transition) was established at the foramen lacerum, bone removal over the ICA proceeded superiorly.

The bone over the anterior genu, horizontal segment, and parasellar carotid protuberance was drilled to eggshell thinness and removed to allow the carotid to be moved laterally without constriction (**Fig. 7.15c**). The medial portion of the clivus at the petroclival junction was drilled down until the lingual process was identified and removed, allowing direct access to periosteum around the ICA. Next, extending the exposure in a rostral direction granted access to the inferior cavernous sinus and quadrangular space and Meckel's cave.

The next landmark is V2. The maxillary antrostomy was widened laterally to expose the posterior wall of the maxilla and isolate this nerve, which was followed superiorly to the foramen rotundum. Bone removal was extended until V2 pierces the dura mater of the middle fossa. The bone between V2 and the vidian canal was also removed toward the junction of the horizontal petrous segment and the anterior genu (**Fig. 7.15d**).

As mentioned previously, the entry into Meckel's cave was through the quadrangular space bounded by the paraclival ICA medially, the horizontal segment of the petrous ICA inferiorly, the dura mater of the middle fossa and V2 laterally, and the abducens nerve superiorly (**Fig. 7.17**). Cranial nerves III, IV, and VI, as well as motor V were monitored during the surgery and a window with no cranial nerve response after stimulation

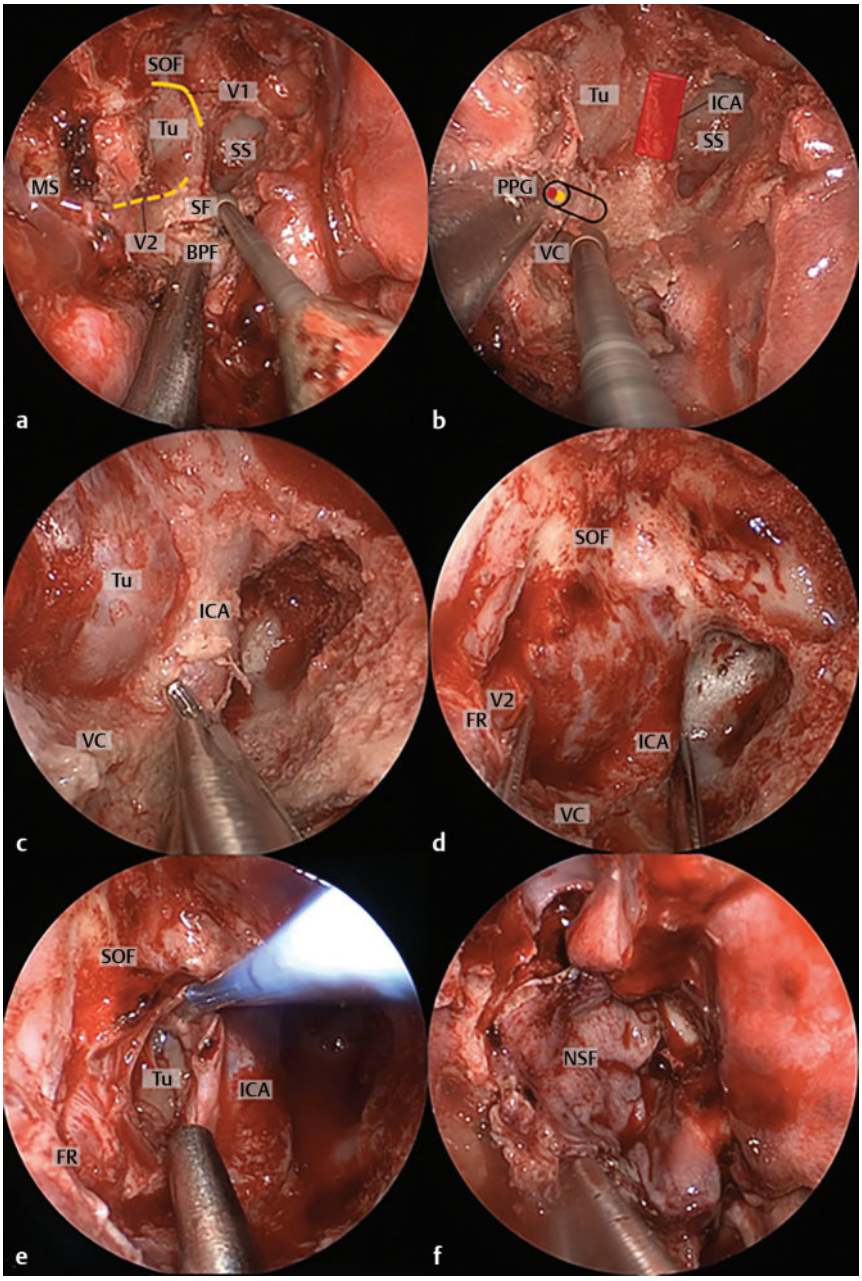


Fig. 7.15 Intraoperative images of the endoscopic endonasal approach for resection of the tumor. (a) After the initial steps, the basopharyngeal fascia was stripped away from the sphenoid floor, which was drilled in a medial-to-lateral progression to identify the vidian canal. Note the tumor bulging into the lateral recess of the sphenoid sinus, displacing V1 and V2 from their normal positions. **(b)** The pterygopalatine ganglion was lateralized and the vidian canal (containing the vidian nerve and artery), once identified anteriorly, was followed posteriorly from its inferior aspect to identify and expose the anterior genu of the ICA, which lies superiorly to the vidian canal. **(c)** Using the vidian canal as a landmark, the anterior genu of the ICA was identified and the bone over it was drilled and removed. **(d)** Once the bone over the carotid was removed, the landmarks were confirmed visually and, with the assistance of nerve stimulation and micro-Doppler, a window with no nerve response was defined. Through this window, we proceeded with tumor resection. **(e)** After dural opening, the tumor can be identified, debulked, and resected protecting the cranial nerves and the ICA. **(f)** Reconstruction of the defect was done using an inlay collagen matrix and the nasoseptal flap. BPF, basopharyngeal fascia; FR, foramen rotundum; ICA, internal carotid artery; MS, maxillary sinus; NSF, nasoseptal flap; PPG, pterygopalatine ganglion; SF, sphenoid floor; SOF, superior orbital fissure; SS, sphenoid sinus; Tu, tumor; V1, orbital division of the trigeminal nerve; V2, maxillary division of the trigeminal nerve; VC, vidian canal.

was defined. The tumor was entered at the level of V2, and the trigeminal nerve was only completely visualized at the end of the resection, since it was laterally positioned. Following the tumor posteriorly, the whole tumor, including the posterior fossa component, was removed with no intraoperative complications (**Fig. 7.15e**).

Reconstruction was performed not only using the nasoseptal flap, but also autologous fat graft was placed protecting the ICA from desiccation and/or trauma (**Fig. 7.15f**).

■ Aftercare

Postoperative examinations confirmed a total resection (**Fig. 7.18**). A few days later, the patient presented with fever and elevated white blood cell count. A CSF sample confirmed high white cells with negative cultures, and the patient received 14 days of intravenous antibiotics. He was discharged with no new neurological deficit on postoperative day 14.

After approximately 8 to 12 months, the facial sensation of the patient normalized and he currently has no complains of facial pain or numbness. Four years having passed since his surgery, the patient has been followed with regular imaging examinations with no signs of recurrence (**Fig. 7.19**).

■ Commentary

After the pioneering work of Jho and Carrau in the mid-90s outlining the tenets of the technique, the use of endoscopy for skull base surgeries, particularly for transsphenoidal pituitary surgery, has steadily grown in the past three decades spurred by a substantial number of anatomical studies, innovations in monitoring techniques, and refinement of fiber-optic

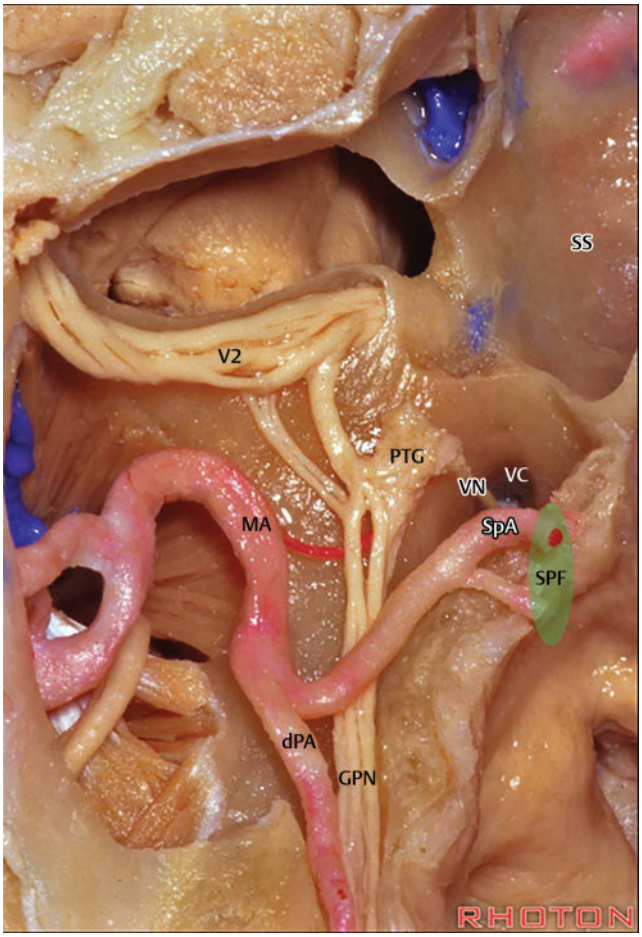


Fig. 7.16 Contents of the right pterygopalatine fossa. The maxillary sinus lies anterior to the pterygopalatine fossa. Removal of the posterior wall of the maxillary sinus in this cadaveric dissection exposes the pterygopalatine fossa. During surgery, the pterygopalatine fossa is exposed after the posterior maxillary antrostomy. The vidian nerve runs in the floor of the lateral recess of the sphenoid sinus. The nerve is followed posteriorly until it meets the lateral aspect of the internal carotid artery at the foramen lacerum. dPA, descending palatine artery; GPN, greater palatine nerve; MA, maxillary artery; PTG, pterygopalatine ganglion; SpA, sphenopalatine artery; SPF, sphenopalatine foramen; SS, sphenoid sinus; V2, maxillary nerve; VC, vidian canal; VN, vidian nerve. (The Rhoton Collection.)

and image-guidance technology. Nowadays, the expanded EEA derived from the continuing collaboration between neurosurgery and otorhinolaryngology has allowed the endoscopic technique to reach an ever-expanding number of lesions and areas of the skull base. These expanded techniques present numerous advantages for different skull base lesions, such as minimal manipulation of the brain, cranial nerves, and blood vessels while offering panoramic exposure of the lesion. It is, on the other hand, a challenging technique with a steep learning curve, requiring dedicated training.

However, it is important to realize that a skull base surgeon must not be a zealot, trying to use the same technique for all patients. A careful analysis of the surrounding anatomy,

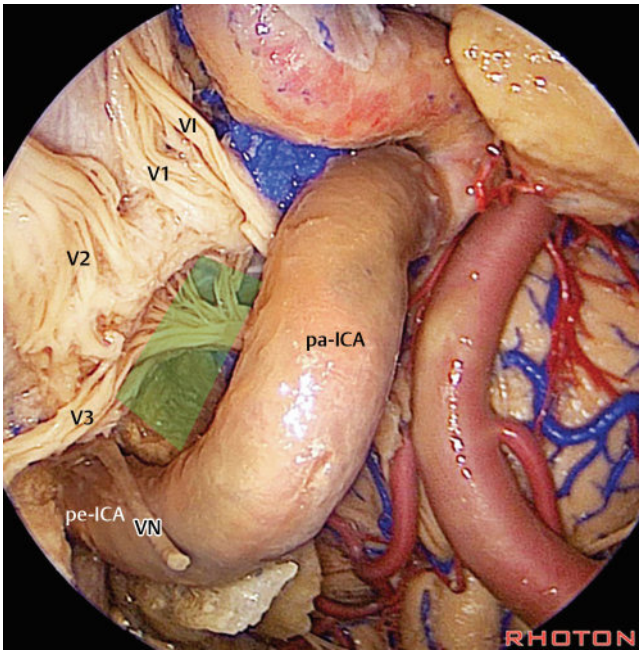


Fig. 7.17 Entry into Meckel's cave. The dura of the middle fossa in this cadaveric dissection has been opened to expose Meckel's cave. The *green shaded area* represents the quadrangular space used for entry into the Meckel's cave. It is bounded the paraclival internal carotid artery medially, the petrous carotid inferiorly, V2 laterally, and the abducens nerve superiorly in the cavernous sinus. Pa-ICA, paraclival carotid artery; pe-ICA, petrous carotid artery; VI, abducens nerve; VN, vidian nerve (The Rhoton Collection.).

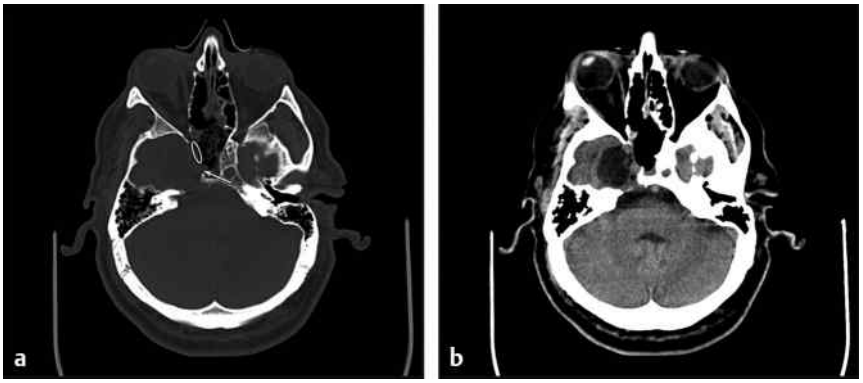


Fig. 7.18 Postoperative CT. **(a)** Bone window. As planned preoperatively, Mullan's triangle has been opened to communicate the lateral recess of the sphenoid sinus and the middle fossa (*oval*), through which tumor resection was done. Note that the osseous encasement of the internal carotid artery has been removed, preserving the vessel (*arrow*). **(b)** Soft tissue window. A complete resection, including the posterior fossa component has been achieved.

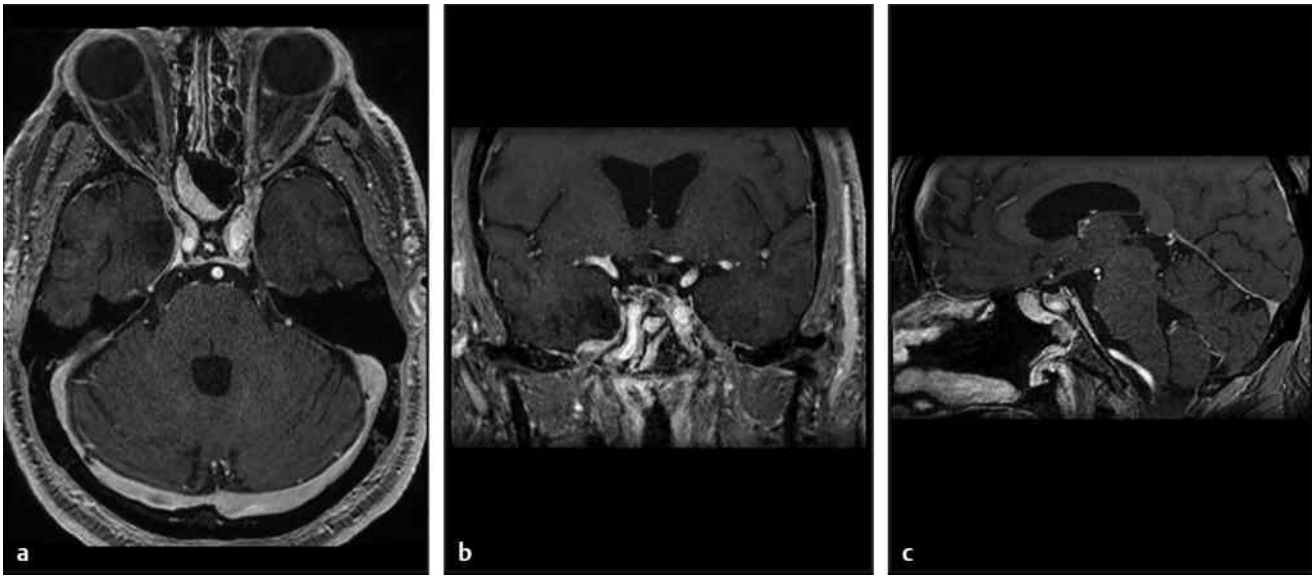


Fig. 7.19 Postoperative MRI at 4-year follow-up. **(a)** Axial, **(b)** coronal, **(c)** sagittal view. The nasoseptal flap used for reconstruction of the defect and protection of the internal carotid artery can be seen enhancing and no signs of tumor recurrence or residual were identified.

particularly relating to the position of cranial nerves and vascular structures is paramount in choosing the safest route to a lesion. Related to trigeminal schwannomas, the EEA is not suitable for tumors blocked anteriorly by the carotid artery, without projection into the sphenoid sinus, with large posterior fossa components. It is, however, uniquely suited for tumors anteromedial to the ICA, occupying the anterior and inferomedial aspect of Meckel's cave. As always, the objectives of the surgery must be clearly defined and the experience of the surgical team carefully assessed, before determining if the EEA is ultimately best for the patient.

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8 Tentorial Incisura

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Keywords: transtentorial, incisural space, vein of Rosenthal, vein of Galen

■ Case Presentation

A 45-year-old woman presented with several weeks of progressive left facial numbness and right arm weakness. Her physical examination was significant for left facial numbness in the V1 and V2 distribution. A subsequent magnetic resonance imaging (MRI) with and without contrast demonstrated a left subtemporal lesion situated along the tentorial incisura in the supra- and infratentorial fossa. There was mass effect on the midbrain leading to effacement of the cerebral aqueduct and subsequent mild triventricular hydrocephalus (**Fig. 8.1**).

Questions

1. What is the most likely diagnosis and how would you classify this tumor anatomically?
2. What is the most likely blood supply to this mass?

■ Diagnosis and Assessment

The MRI revealed a homogenously enhancing, extra-axial mass compressing the upper brainstem, with the medial pole of the tumor reaching in the midline. On first glance, one might assume this mass to be attached to the petrous ridge, but in fact it is based along the tentorium and has a dural tail (**Fig. 8.2**). With the degree of brainstem compression, it was surprising that the patient complained only of right arm weakness and a numb face, but the lack of other symptoms hinted at a likely slow pace of tumor growth. In the superior/inferior dimension, the tumor extended from midpoint of third ventricle (level of the massa intermedia) to the upper part of the posterior fossa. Laterally, it appeared to reach the petrous ridge near the superior petrosal sinus.

The imaging findings are typical for a meningioma originating from the tentorial incisura. The differential diagnosis for dural-based tumors includes hemangiopericytomas, solitary fibrous tumors, sarcoidosis, and dural-based metastasis. Although it may be difficult to distinguish other dural-based lesions from meningiomas on imaging alone, clinical history and subtle radiographic findings can be useful. Meningiomas are typically slow growing and therefore have an insidious onset of symptoms. The lack of history of malignancy in this patient makes a dural-based metastasis unlikely, and the absence of bony destruction or flow voids within the lesion renders hemangiopericytoma unlikely as well.

Despite the vast surface area of the tentorium, covering the cerebellum and separating the supra- and infratentorial compartments, tentorial meningiomas represent only 3 to 6% of all intracranial meningiomas. As the tentorium invests along the internal occipital protuberance posteriorly and attaches to the petrous ridge anteriorly with dural folds extending to the anterior clinoid process, tentorial meningiomas refer to an anatomically heterogeneous group of extra-axial tumors that may arise in either the posterior or middle cranial fossa. As such, meningiomas of this region may lead to a variety of neurological manifestations, which may make their initial diagnosis challenging. Indeed, the delayed detection of these tumors has been proposed by some authors to account for their often large size at initial diagnosis. Tentorial meningiomas have been classified by Yasaragil by their occurrence relative to the inner ring ("free edge") or the outer ring (along the transverse sinus) of the tentorium. Meningiomas of the inner ring may arise from the tentorial apex (at the confluence of the vein of Galen and the straight sinus) or along the tentorial incisura. Tentorial meningiomas of the "outer ring," meanwhile, can occur along the falcotentorial junction and transverse/sigmoid sinus, adjacent to the torcula.

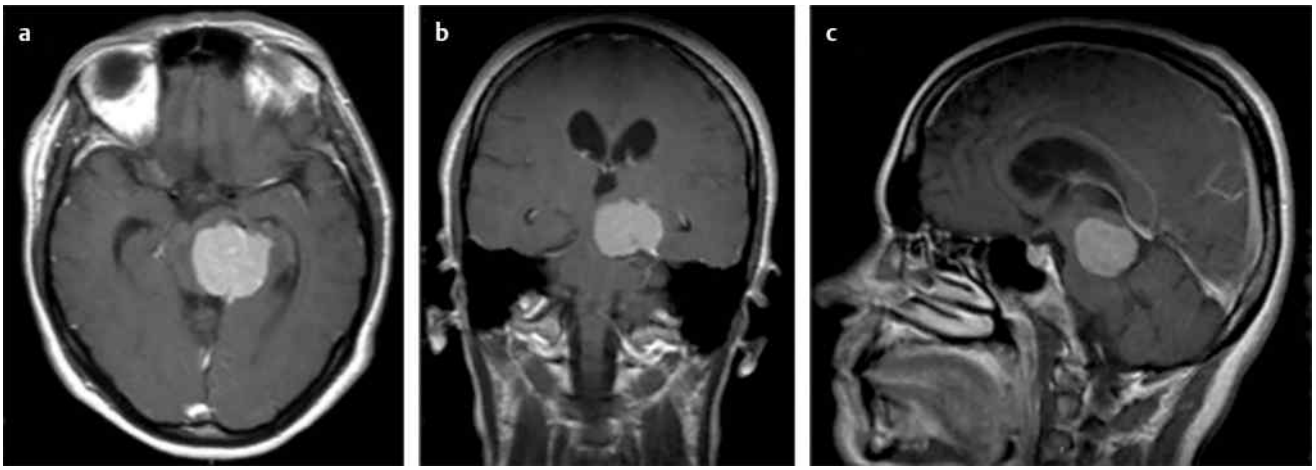


Fig. 8.1 **(a)** Axial, **(b)** coronal, and **(c)** sagittal T1-weighted postcontrast magnetic resonance images depicting a contrast-enhancing mass situated along the medial edge of the tentorium with both supra- and infratentorial extension. There was mass effect on the midbrain with resultant mild hydrocephalus.

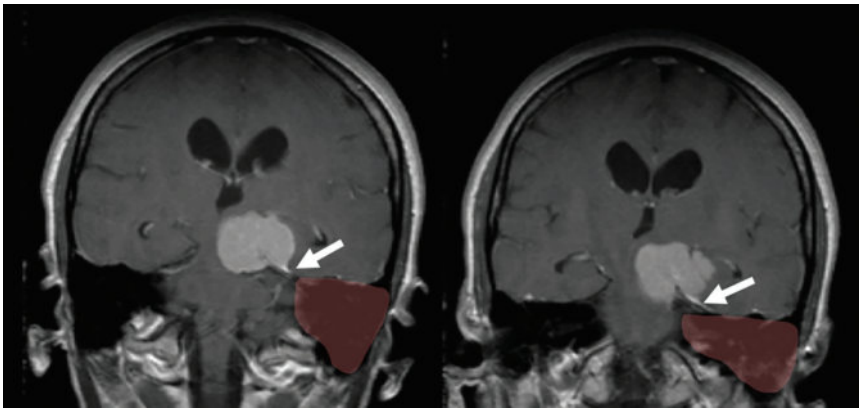


Fig. 8.2 Preoperative anatomical assessment. Coronal views confirmed that the tumor was attached to the tentorium and entirely separate from the petrous apex. *Pink shaded area*, petrous temporal bone; *white arrow*, gap between tumor and petrous apex and dural tail.

Alternatively, they may reside in paramedian locations. Tentorial meningiomas may be based on either the superior or inferior face of the tentorium and therefore may reside within the supratentorial compartment, the infratentorial space, or have extensions in both compartments.

In this case, the patient had a meningioma with both supra- and infratentorial extension situated along the tentorial incisura, the triangularly shaped inner ring of the tentorium. Rhoton subdivided the incisura into three anatomical spaces: (1) a single anterior incisural space (AIS) (anterior to the brainstem), (2) a paired middle incisural space (MIS) (lateral to the midbrain), and (3) a single posterior incisural space (PIS) (posterior to the brainstem) (**Fig. 8.3**). Samii recognized that meningiomas of the incisura typically occupy either the middle or PISs, and according to his classification, this patient's tumor occupies the MIS.

As this patient presented with progressive symptoms from a benign disease with no significant comorbidities, surgical intervention was the only logical management. The goal of surgery was primarily to decompress the brainstem and prevent worsening hydrocephalus, and secondarily to remove the tumor as much as possible without generating neurological deficits. The hope was to resect the tumor completely to minimize the chances of recurrence in this young patient. She was admitted to the hospital for close neurological monitoring and in the absence of peritumoral edema, anticonvulsants and steroids were deferred prior to surgery. A magnetic resonance venography (MRV) was obtained in order to delineate the deep venous drainage around the tumor and the course of the vein of Labbé for subsequent surgical planning. The main blood supply to tentorial meningiomas is usually from the artery of Bernasconi and Cassinari (branch of the meningohypophyseal trunk), a marginal tentorial branch of the inferolateral trunk, and meningeal feeders off of the posterior cerebral artery/superior cerebellar artery (PCA/SCA). Selective embolization of these branches is difficult, and therefore preoperative tumor embolization was not performed in this case.

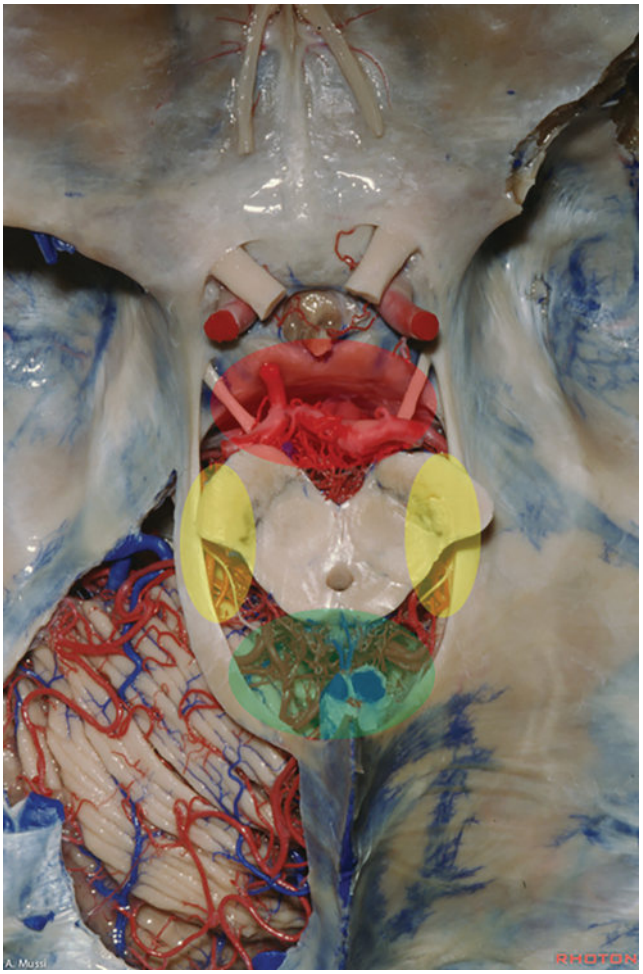


Fig. 8.3 The tentorial incisura. The area between the midbrain and the free edges is divided into an anterior incisural space located in front of the brainstem and anterior wall of the third ventricle (red); paired middle incisural spaces situated lateral to the midbrain (yellow); and a posterior incisural space located behind the midbrain (green). The middle incisural space is the site of the crural and ambient cisterns; and the posterior incisural space is the site of the quadrigeminal cistern. (The Rhoton Collection.)

■ Anatomical and Therapeutic Considerations

The outer ring of the tentorium invests along the petrous portion of the temporal bone (enclosing the superior petrosal sinus) and the internal occipital protuberance (enclosing the transverse sinus and torcula). From these attachments, the tentorium is angled upward to its apex where it attaches to the falx, forming the falcotentorial junction that encloses the straight sinus. The tentorial incisura is the triangular opening along the tentorium and is the only portion of the tentorium without a direct osseous attachment. As mentioned above,

meningiomas of the tentorial incisura tend to grow into either the middle or PIS.

The MIS occupies the region between the free edge of the tentorium and the brainstem and is intimately related to the medial temporal lobe, deep venous drainage, and the posterior circulation. The free edge of the tentorium lies at the level of the pontomesencephalic sulcus, which separates the midbrain and pons and represents the medial border of the MIS. The medial surface of the temporal lobe, containing the uncus and parahippocampal gyri, forms the lateral boundary of the MIS. The optic tract and medial and lateral geniculate bodies form the roof. The supratentorial part of the MIS contains the crural cistern, located between the cerebral peduncle and the uncus, and the ambient cistern, which is continuous posteriorly with the quadrigeminal cistern.

The major arteries in the MIS are the anterior choroidal, PCA and SCA (**Fig. 8.4**). The anterior choroidal artery courses just superior to the free edge of the tentorium, while the PCA travels

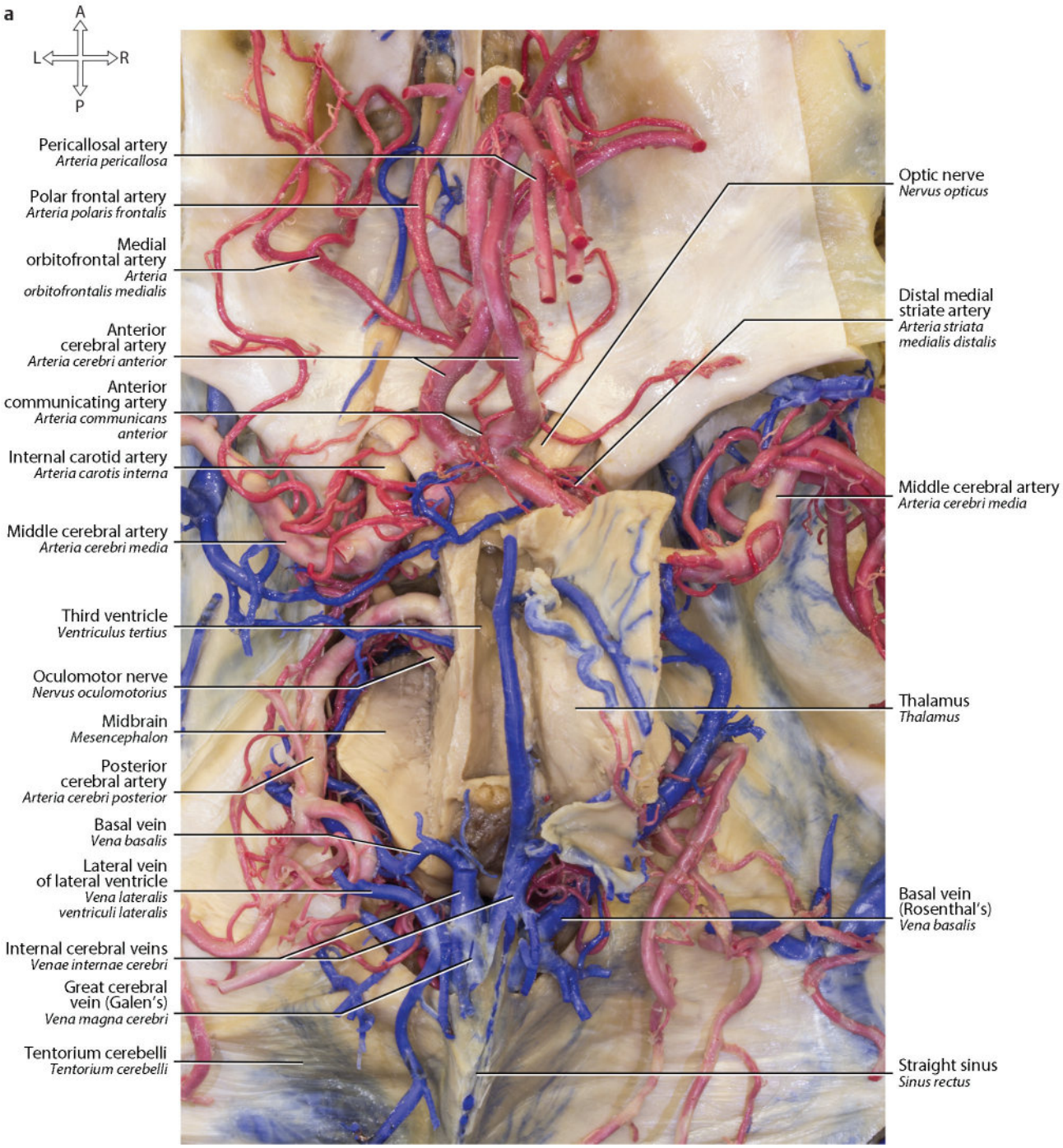


Fig. 8.4 Superior view of the tentorial incisura. (a) The major arteries in the middle incisural space are the anterior choroidal, posterior cerebral (PCA), and superior cerebellar arteries (SCA).

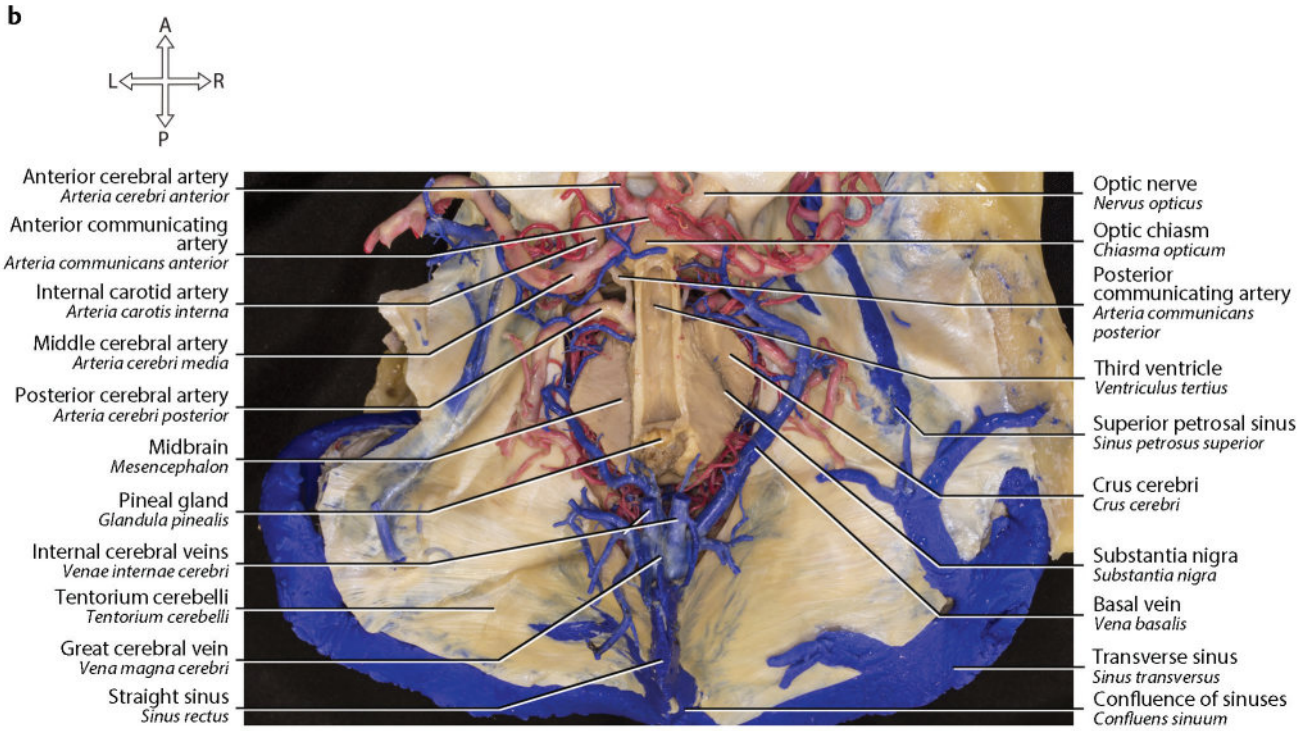


Fig. 8.4 *(Continued)* **(b)** The basal vein of Rosenthal courses through the ambient cistern within the middle incisural space. (Reproduced from Peris-Celda M, Martinez-Soriano F, Rhoton AL Jr., eds. Rhoton's Atlas of Head, Neck, and Brain. 2D and 3D Images. 1st ed. Thieme; 2017.)

through the crural and ambient cistern at the level of the tentorial edge, giving off several cortical branches that will cross the free edge of the tentorium en route to the inferior temporal lobe. Other branches of the PCA residing in the MIS include the medial and lateral posterior choroidal arteries that supply the choroid plexus in the third ventricle and atrium, respectively. The thalamogeniculate artery, another branch of the PCA found within the ambient/crural cistern, supplies the thalamus and geniculate bodies. The superior cerebellar artery travels beneath the level of the tentorial edge. The venous drainage within the MIS is predominantly through the basal vein of Rosenthal, which eventually drains into the vein of Galen in the quadrigeminal cistern.

Both the cranial nerves V and IV are closely related to the middle incisural region. The trochlear nerve exits the brainstem in the quadrigeminal cistern just beneath the inferior colliculus prior to traveling between the PCA and SCA in the MIS. At the level of the tectum, cranial nerve IV remains medial to the free edge of the tentorium, but at the level of the peduncle, it follows the free edge prior to piercing the oculomotor triangle. Prior to entering the cavernous sinus, cranial nerve IV is ensheathed within the anterior petroclinoid fold, the anterior extension of the tentorium that invests on the anterior clinoid process. The trigeminal nerve is situated beneath the free edge of the tentorium and courses through the infratentorial portion of the MIS prior to entering Meckel's cave.

Options for Approach

On first inspection of the MRI (**Fig. 8.5**), it was easy to confuse this with a petroclival meningioma, the removal of which might require a posterior petrosectomy. On closer inspection, it becomes clear that this tumor is entirely separate from the middle fossa floor and petrous apex. Years having passed since this patient's story unfolded, we now have technology that can efficiently render two-dimensional preoperative images into three dimensions (**Fig. 8.5**). These pictures once again confirm that the tumor was separate from the temporal bone and appears to "float" above the petrous ridge, and also show us that this patient's tumor was approximately two-thirds above and one-third below the tentorium. Since this tumor had a split existence in the middle and posterior fossa, logically, the options for approach are supratentorial, infratentorial, or combined.

An infratentorial-only approach, using the posterolateral corridor via a retrosigmoid opening, has a major advantage of avoiding any retraction on the dominant temporal lobe. Gravity-assisted retraction of the cerebellum can provide sufficient exposure of the paramedian supracerebellar corridor, and by following the inferior surface of the tentorium, the posterior fossa component of the tumor can be safely accessed. Subsequent coagulation and incision of the tentorium turn this into the supracerebellar transtentorial (SCTT) approach, and the supratentorial component can be resected via this approach. Although the corridor may seem small, even large tumors above the tentorium can be resected this way, as the coagulation of the tentorium devascularizes the tumor, and the ensuing tumor removal progressively enlarges the intradural surgical window. It is intuitive to assume that the strength of an infratentorial approach lies in handling the posterior fossa contents. However, because of the inferior-to-superior surgical trajectory, the SCTT approach actually provides better access to the rostral/medial tip of the tumor *above* the tentorium than supratentorial approaches, as the latter requires massive

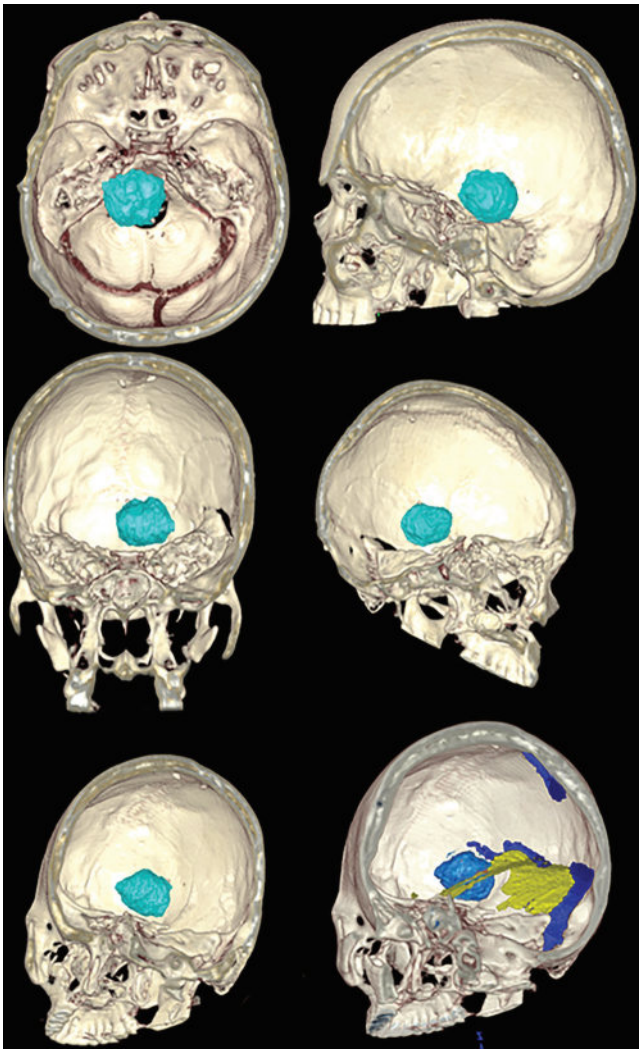


Fig. 8.5 Three-dimensional rendering of the tentorial tumor (aqua blue) relative to petrous ridge. Note that the tumor "floated" about the petrous temporal bone as it was attached to the tentorium and not the petrous bone. The tentorium is rendered in yellow, and viewing the tumor along the plane of the lateral tentorium, it appeared two-thirds above and one-third below the tentorium.

retraction forces to visualize the same parts from under the temporal lobe. The disadvantage of this technique is that the working distance to the target is quite long, and maneuvering around the brainstem and tentorium, especially one that is hypervascularized, at such a distance is technically challenging.

In contrast to the SCTT approach, a short working distance and straight trajectory are the major advantages of the subtemporal approach. Since the majority of our patient's tumor is above the tentorium, expedient debulking can enlarge the surgical window quickly, which in turn makes coagulation and incision of the tentorium, and devascularization of the rest of the tumor more efficient. Another potential advantage of this approach is that if any part of the tumor is adherent to the brainstem, the wider, straighter corridor would significantly facilitate microsurgical dissection of these adhesions. However, the major disadvantage is that adequate exposure of the tentorium incisura through the subtemporal corridor requires retraction of the temporal lobe, with the associated risk of injury to the vein of Labbé as well. Both retraction as well as venous injury can cause harm to the dominant temporal lobe in this patient.

Recently described, the transzygomatic approach with inferior temporal gyrectomy was designed specifically to mitigate the retraction on the temporal lobe and risk to the vein of Labbé. However, by moving the starting point of the surgical trajectory more anteriorly, the distance to target is once again quite long and almost comparable to the SCTT approach. Another option to avoid temporal lobe retraction is the aforementioned posterior petrosectomy. By removing the bone along the petrous ridge, the superior petrous sinus can be coagulated and incised. In turn, the tentorium can then be cut from a lateral-to-medial direction, and both the supra- and infratentorial portions of the tumor can then be removed via the posterolateral corridor (**Fig. 8.6**). In this option, the surgical corridor is expanded by removing the bone of the petrous ridge, thus obviating the need for temporal lobe retraction. However, the posterior petrosectomy significantly extends the operative time, and its advantages may be nullified by this.

Approach of Choice

In the end, we designed a bone opening that would give us access to both the lateral subtemporal, as well as the paramedian

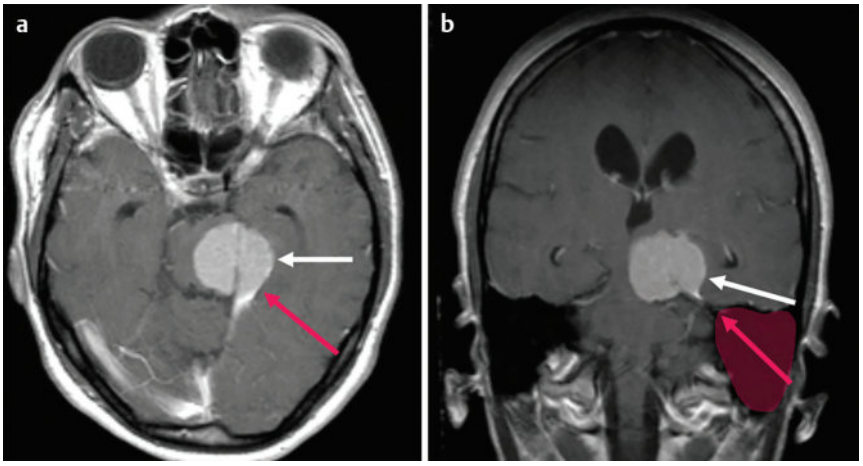


Fig. 8.6 Comparing the lateral and posterolateral corridors. (a) Axial and **(b)** coronal preoperative images showing both surgical trajectories. *White arrow*, the lateral corridor and the subtemporal approach; *pink arrow*, the posterolateral corridor and the posterior petrosectomy approach. Although the petrosectomy can minimize the retraction needed on the temporal lobe, it would extend the duration of surgery significantly in this case.

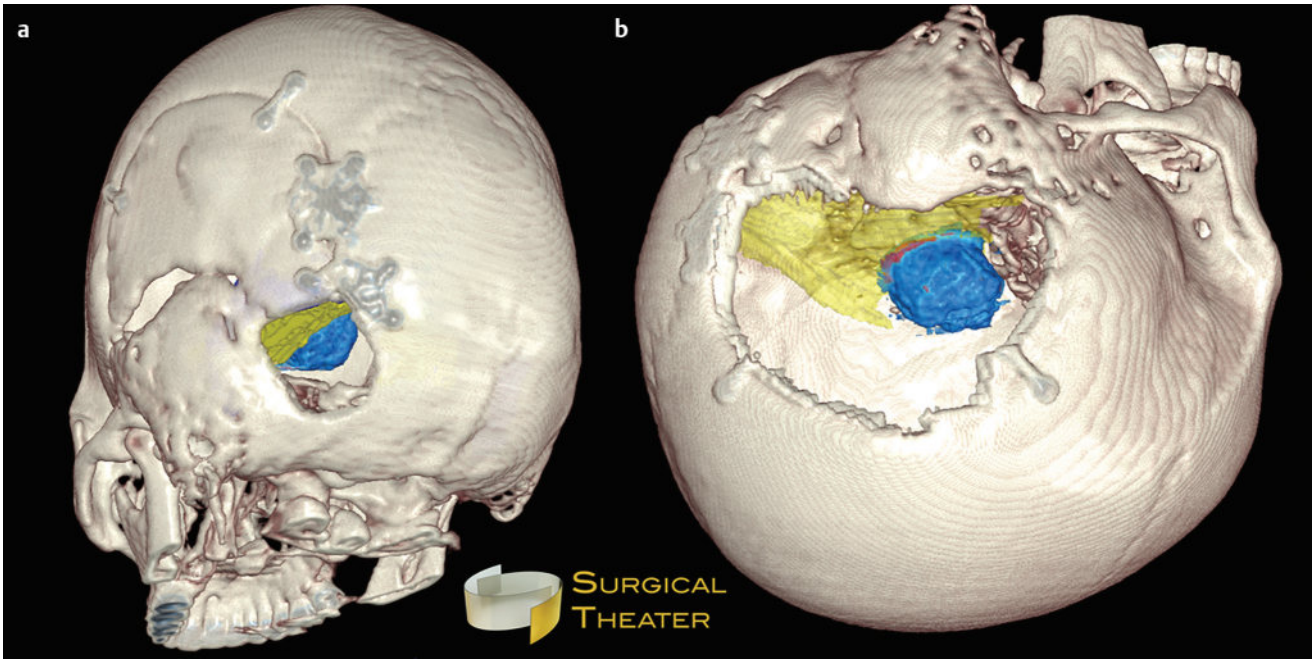


Fig. 8.7 Three-dimensional renderings. Preoperative images of the patient's tumor are superimposed on postoperative images of her bone opening. **(a)** The patient's tumor (*blue mass*) as viewed from the retrosigmoid bone opening, along the inferior surface of the tentorium; **(b)** her tumor as viewed from the temporal bone opening, along the superior surface of the tentorium.

supracerebellar corridors. Since the majority of the tumor was supratentorial, our intent was to first use the shorter, straighter subtemporal corridor to expeditiously debulk and devascularize the tumor. However, as our patient's tumor extended quite high, with the superior pole reaching approximately 2 cm above the middle fossa floor, we feared that excessive dominant temporal lobe elevation would be necessary to reach the most rostral and medial portions of the tumor. As such, we prepared to perform a concurrent left-sided paramedian suboccipital craniotomy to access the medial and infratentorial portions of the tumor through a paramedian supracerebellar approach. Thus, a combined subtemporal, SCTT approach was the surgical scheme.

With surgical planning technology that is a decade in advance of this patient's operation, we are now able to see the tumor in three-dimensional renderings through the patient's actual operative windows, and compare the vantage point from the paramedian supracerebellar corridor and the lateral subtemporal corridor (**Fig. 8.7**).

Questions

1. How would you position the patient and design the incision for this combined approach?
2. What measures would you take to protect the left transverse sinus during surgery?
3. Can you think of other protective measures that can potentially mitigate the risk of temporal lobe retraction?

Description of the Technique

After the patient was under general anesthesia, a lumbar drain was placed to facilitate brain relaxation during surgery. The patient was then positioned laterally with the head turned rightward in order to gain access to both the suboccipital region and the middle cranial fossa floor. As the subtemporal corridor was our primary approach to the target, the root of the zygoma was positioned at the highest point (vertex angled to the floor approximately 20 degrees) in order to facilitate gravity-assisted mobilization of the temporal lobe. During positioning, we avoided excessive rotation of the neck, which could lead to occlusion of the jugular veins and increased intraoperative bleeding (see **The Three Approach Elements**).

An upside-down "U-shaped" incision was used, starting in front of the left tragus, moving up, then posteriorly, and then down the suboccipital region with a curve toward the mastoid tip. The temporalis was reflected inferiorly. Four burr holes were made, two on either side of the transverse sinus, and a high-speed drill was then utilized to connect them, creating an "L-shaped" bone flap with the long limb along the squamous

The Three Approach Elements

Corridor: subtemporal

Craniotomy: middle fossa/temporal

Modifier: tentorial incision

temporal bone. The osteotomies across the transverse sinus were made with the Kerrison rongeurs. After removal of the "L-shaped" bone flap, the squamous temporal bone was shaved so that it was flush with the middle fossa. Dural opening was made again as an upside-down "U," flapped inferiorly along the middle fossa floor, and the posterior fossa dura was untouched for now. The temporal lobe was elevated as spinal fluid was released from the lumbar drain. Using the navigation system, the tumor was located at the tentorial incisura under the temporal lobe. Tumor debulking was undertaken using ultrasonic aspirator (see **Operative Setup**).

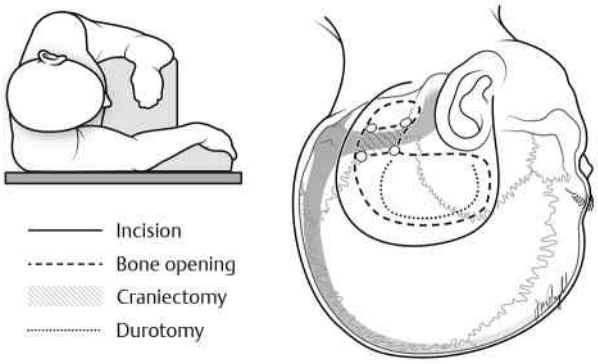
Operative Setup

Position: lateral, left side up

Incision: upside-down "U"

Bone opening: "L-shaped" subtemporal/retrosigmoid

Durotomy: upside-down "U" over temporal lobe



By stepwise debulking, the tumor capsule was gradually dissected free from the brainstem and was completely resected. During the process, the tentorium was coagulated and incised thereby opening an aperture into the posterior fossa (**Fig. 8.8**). The trochlear nerve was seen underneath the tumor and unharmed. In this case, adequate bony removal and judicious use of lumbar drainage enabled complete removal of the tumor from a subtemporal approach. The supracerebellar corridor was left unused.

Can one argue that it was a "complication," opening a corridor that went unused? For us, it was a matter of added safety. There exist no preoperative tests that can predict with complete certainty whether the rostromedial pole of the tumor would "peel" off the brainstem and roll out smoothly. In this patient, it did, but had it been adherent to the brainstem, the inferior-to-superior view from the infratentorial corridor would have been useful to facilitate safe dissection of this portion despite a long reach.

Surgical Pearls

1. When performing a subtemporal approach to the tentorial incisura, temporal lobe elevation is unavoidable. Use of lumbar drainage is imperative to facilitate brain relaxation and to prevent excessive retraction on the temporal lobe. Similarly, positioning the head such that the vertex is oriented inferiorly is one way to facilitate gravitational (not mechanical) elevation of the temporal lobe.
2. The vein of Labbé must be preserved during the subtemporal approach. Injury to the vein of Labbé can lead to a temporal lobe venous infarction. Identifying the vein on preoperative imaging is of paramount importance. Dural opening, and placement of retractor, should be tailored to avoid injury of this vein.
3. Opening the tentorium through the subtemporal approach can allow access to the posterior fossa and may be necessary to resect incisural meningiomas extending into the infratentorial space. Care must be taken to avoid injury to the fourth nerve given its course adjacent to the free edge of the tentorium.

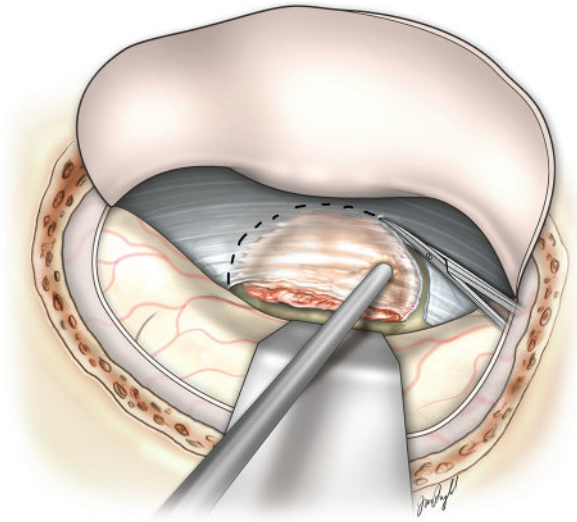


Fig. 8.8 Intraoperative view. The left temporal lobe was elevated to show the tumor along the superior surface of the tentorium. The tentorium was incised to provide access to the tumor's posterior fossa/infratentorial component.

Aftercare

The patient experienced difficulty with language, specifically comprehension, following surgery. She was managed with a rapid taper of dexamethasone and was maintained on anticonvulsants for 7 days given the need for temporal lobe elevation. The patient was discharged home on postoperative day 5, and at 3-month follow-up, postoperative MRI showed gross total tumor resection (**Fig. 8.9**). As a professional editor, her dysfunctional comprehension kept her out of work for quite some time. Fortunately, with speech therapy, her language function recovered completely, and she was able to return to work. Final pathology returned as a meningioma WHO grade I.

Five years after her surgery, a surveillance MRI detected recurrence of the tumor along the tentorial edge. Since this was very small, we elected to treat the recurrent tumor with radiosurgery. A 3.5-cc volume was treated to 3,000 cGy in five

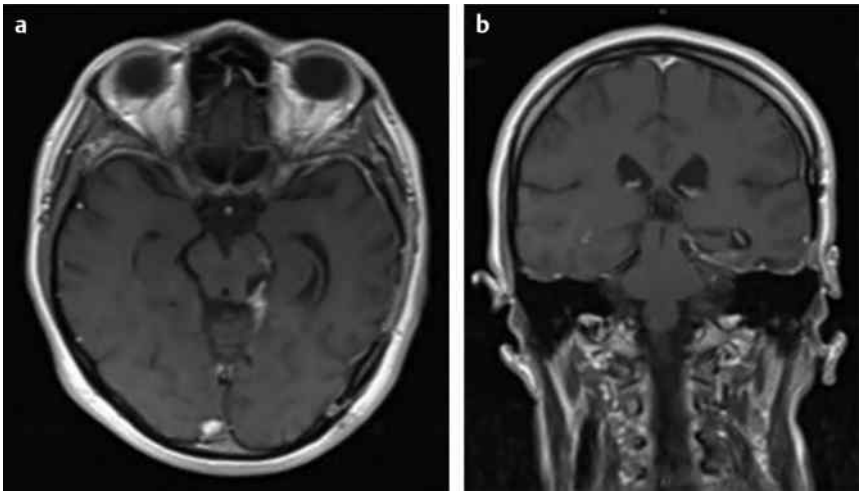


Fig. 8.9 Postoperative images. (a) Axial and **(b)** coronal T1-weighted postcontrast magnetic resonance images after the operation depicting total resection of the tentorial incisural meningioma. The mass effect on the brainstem had resolved.

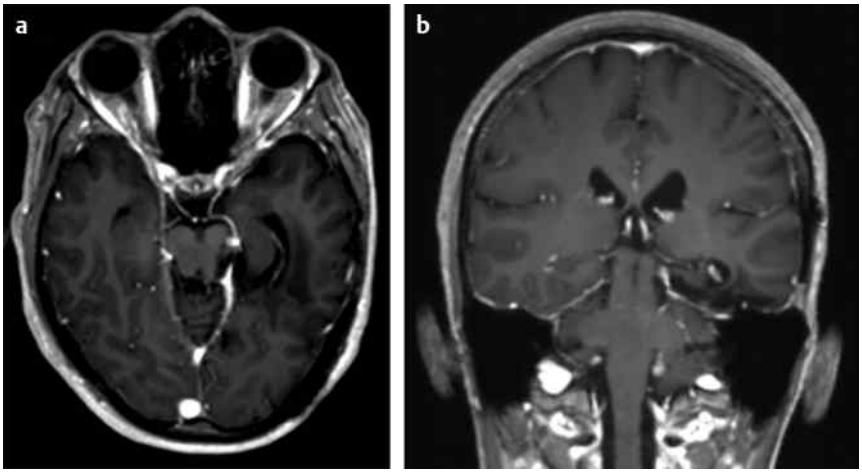


Fig. 8.10 Postoperative images at 10 years. (a) Axial and **(b)** coronal T1-weighted postcontrast magnetic resonance images 10 years after the operation. The tumor had recurred after 5 years and the small recurrence was treated with radiosurgery. Five years after that, no tumor was visible at the original site.

sessions. Another 5 years have passed, and her tumor is barely visible at the time of writing (**Fig. 8.10**).

■ Possible Complications and Associated Management

As with any craniotomy, postoperative hemorrhage, seizure, or infection are possible postoperative complications. The most dreaded complication following a subtemporal craniotomy is injury to the vein of Labbé, which can lead to a temporal lobe venous infarct with resultant hemorrhage, seizure, and poor neurological outcome. Preoperative venograms (such as MRV or CTV) are useful at evaluating the vein, as it courses over the temporal lobe prior to draining into the transverse sinus–sigmoid sinus junction. Such venograms are useful at modifying the trajectory through the subtemporal corridor to mitigate the risk to the vein. Prolonged or excessive elevation of the temporal lobe can also lead to thrombosis of the vein and subsequent venous infarction. Thus, strategies geared toward minimizing temporal lobe retraction (lumbar drainage, head positioning) are necessary. In certain cases, if the vein of Labbé directly traverses the necessary route to the tentorial edge, a different approach may be needed.

Any degree of brain retraction may lead to subsequent edema and damage. In the dominant temporal lobe, this may result in language deficits, as experienced by our patient. Furthermore, excessive cortical retraction may also lead to the development of postoperative seizures. This is especially concerning when elevating the medial temporal lobe, which is necessary in this operation. Prophylactic and short-term anticonvulsants are often administered to patients, although specific evidence supporting this is sparse. Postoperative cerebrospinal fluid leak is another possible complication following a subtemporal approach. Mastoid air cells, if transgressed, must be thoroughly waxed on initial exposure and prior to closure. Any dural defect should be adequately closed either primarily or with dural allograft. In cases of large dural defects, fat grafts or muscle grafts can be considered.

Perspective

Omer S. Sahin, Ulas Cikla, and Mustafa K. Baskaya

■ Introduction

Incisural meningiomas are the most challenging types of the tentorial meningiomas because of their deep location and complex neurovascular relations. The tentorial incisura, or tentorial notch, provides the only connection between supra- and infratentorial compartments, and is the only part of the tentorium that is not attached to the skull. Understanding the complex anatomy of the tentorial incisura and its neurovascular relationships are essential for approaching tentorial tumors and other lesions of the region, including aneurysms, arteriovenous malformations, and dural arteriovenous fistulas.

■ Case Presentation

A 51-year old woman presented with nonspecific speech and walking difficulties over the prior 3 months. The patient also reported having abnormal sensations in her right eye. On neurological examination, she exhibited no focal motor, cranial nerve, or sensory deficits. She had a mild dysmetria on the left side. MRI scan of the head revealed a large homogeneously contrast-enhancing lobulated mass located in the infratentorial region. The mass was approximately 4 × 5 cm and was associated with vasogenic edema in the surrounding brain. There was also a significant mass effect on the brainstem and left cerebellum (**Fig. 8.11**).

■ Anatomical and Therapeutic Considerations

As briefly mentioned in the preceding section, the tentorial incisura can be divided into three compartments, according to its relationship with the brainstem (**Fig. 8.3**). The AIS is located anterior to the pons and midbrain, and extends from the lamina terminalis to the interpeduncular fossa. The cranial nerves that pass through the AIS are the optic and oculomotor nerves. Also located within the AIS are the posterior communicating artery, the anterior choroidal artery, the medial posterior choroidal artery, and the basilar bifurcation and interpeduncular cistern. The AIS also contains the basal vein of Rosenthal that originates below the anterior perforated substance and courses through AIS, MIS, and PIS, before draining into the vein of Galen.

The MISs, one on each side, are situated lateral to the brainstem. Superiorly, the MIS is a narrow space between the midbrain and temporal lobe, and inferiorly, it is between the upper brainstem and cerebellum. The inferior surface of the thalamus is the roof of the MIS, and the lateral wall is composed of the hippocampal formation on the medial side of the temporal lobe. The medial wall is formed by the upper pons and lateral surface of the mesencephalon. The MIS contains the crural and ambient cisterns, the trochlear and trigeminal nerves, and the PCA, SCA, and anterior choroidal artery. Branches of the PCA, including the thalamogeniculate artery and lateral posterior choroidal arteries, course medial to the free edge of the tentorium.

The PIS lies between the posterior edge of the midbrain and the tentorial apex and can be considered to have a roof, a floor, and anterior and lateral walls. The crura of the fornix, the hippocampal commissure, and the lower surface of the splenium together form its roof. The anterior wall of the PIS is formed mainly by the quadrigeminal plate. The cerebellum, vermis, and the quadrangular lobule of the cerebellar hemisphere form the floor, and lastly, both the pulvinar, the crus of the fornix, and the medial surfaces of the hemispheres comprise the lateral wall. The PCA and SCA course within the PIS. The internal cerebral vein and the basal vein of Rosenthal join together in the PIS and become the vein of Galen.

Our patient's MRI showed a lobulated, contrast-enhancing, extra-axial mass, centered on the left tentorium, that was consistent with a meningioma. There was a mass effect on the left cerebellar hemisphere, brainstem, and the confluence of the vein of Galen and straight sinus. The fourth ventricle was compressed, but there was no hydrocephalus (**Fig. 8.11**). Although the patient

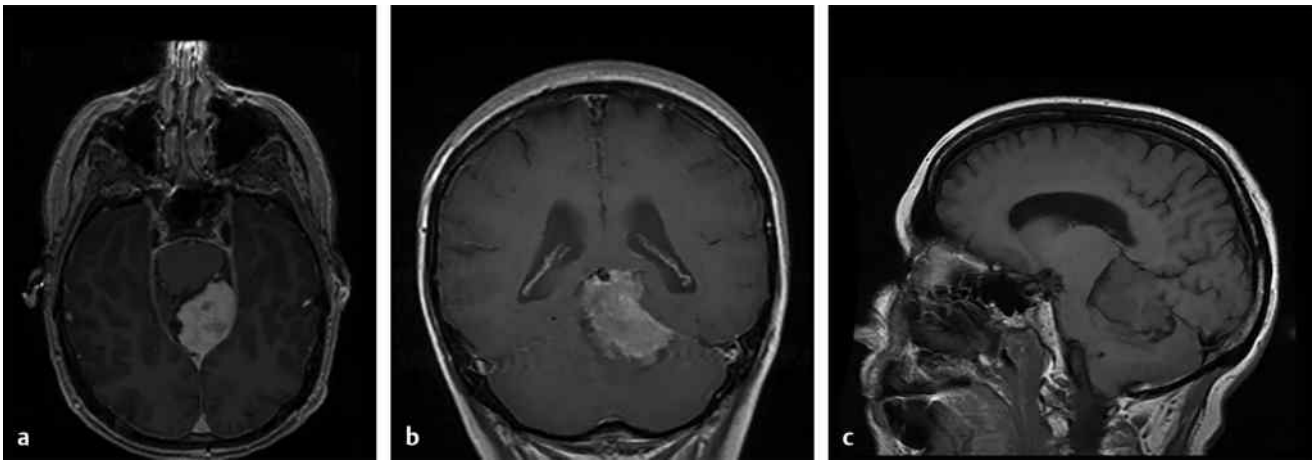


Fig. 8.11 Preoperative images. (a) T1 postcontrast axial and **(b)** coronal MRI showed a large tentorial meningioma in the posterior incisural space. **(c)** T1 sagittal noncontrast MRI showed compression on the midbrain and cerebellum.

had only mild dysmetria on the left, the edema in the surrounding brain prognosticated that this would soon worsen and other symptoms appear. Since the patient was young and healthy, the goal of surgery was set for maximal safe resection.

Options for Approach

The pterional approach is the "workhorse" for approaching various lesions located in the AIS. Since the AIS includes a plethora of arterial branching points, aneurysms are very common in the AIS, and surgery here most often involves these lesions. The second most commonly used approach to access lesions in the AIS is the subtemporal approach.

Surgeries involving the MIS are most commonly done for neoplasms such as gliomas of the mediobasal temporal lobe, thalamic tumors, meningiomas, and Meckel's cave tumors. The most common approaches to MIS lesions are the pterional and subtemporal approaches or a combination of both, the so-called "half-and-half approach." For lesions involving the ambient cistern or cerebellomesencephalic fissure, the surgeon may choose to approach laterally via the paramedian SCTT approach, as this obviates the need for any retraction on the temporal lobe. This is especially worth considering with a lesion on the left side, like the one in the first section of this chapter, when the dominant temporal lobe is involved.

The most common lesions of the PIS are meningiomas arising from the infra- or supratentorial aspect of the tentorium, falcotentorial meningiomas, pineal tumors, and gliomas. When considering the PIS, there are four different types of approach that are used according to the location of the tumor: (1) the supracerebellar infratentorial (SCIT) approach with its modifications such as the paramedian and transtentorial approaches, (2) the occipital transtentorial approach, (3) the posterior interhemispheric approach, and (4) the combined SCIT with occipital approach.

The SCIT is usually the approach of choice to access the lower half of the PIS in the midline or near-midline. It is excellent for tumors with a major component below the tentorium, impinging on the apex of the cerebellum and quadrigeminal plate. If the tumor originates above the vein of Galen, the occipital transtentorial or posterior interhemispheric approach should be considered. For intrinsic brain tumors located in the region of the isthmus, involving the posterior parahippocampal and lingual gyri, the occipital interhemispheric approach can be selected. Furthermore, for these locations, the SCTT approach should also be in the surgeon's armamentarium. Finally, for very large tumors involving both infra- and supratentorial compartments, a combination of supracerebellar and occipital approaches have also been used by various neurosurgeons.

Approach of Choice

Returning to our patient, the tumor was located mainly in the PIS, with an extension into the left middle incisural space. The bulk of the tumor was infratentorial, but again, there was a small extension above the tentorium. Since the major component was below the vein of Galen, the decision was made to approach the tumor via the SCIT approach, with a transtentorial extension to resect the supratentorial portion **(Fig. 8.12).**

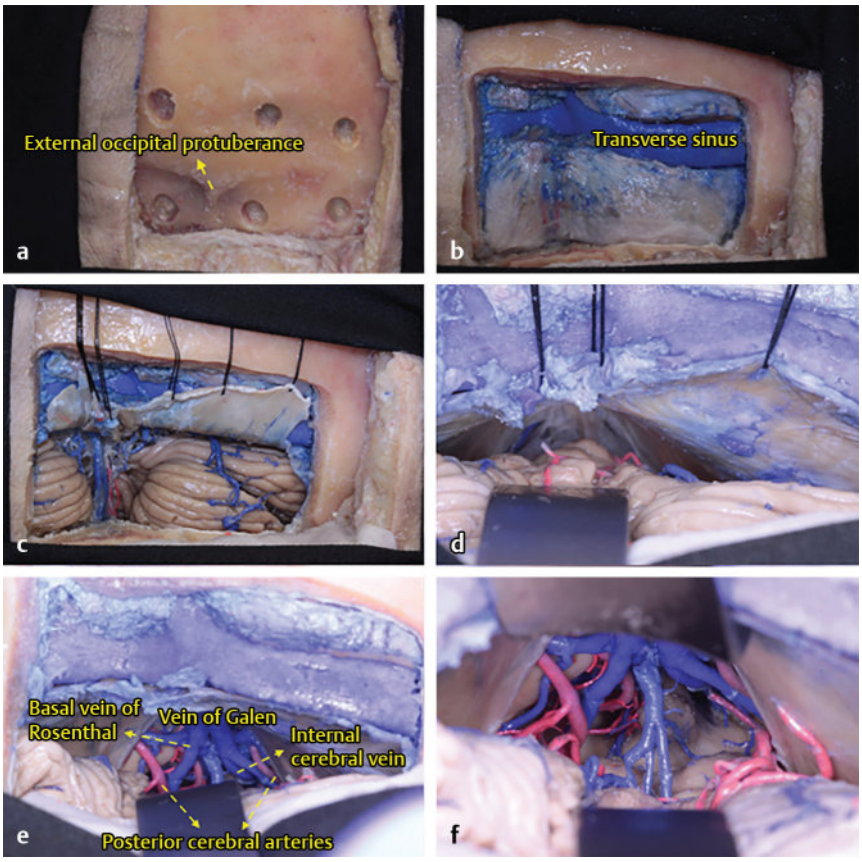


Fig. 8.12 Cadaveric dissection pictures that simulate the midline supracerebellar infratentorial approach. Multiple burr holes have been placed on both sides of the transverse and sagittal sinuses for safe elevation of the dura **(a)**. Exposure after elevating the bone flap. Note that craniotomy is eccentric to left side **(b)**. Dural opening and retraction of the dura with sutures over the transverse sinus **(c, d)**. Vascular structures observed include the basal vein of Rosenthal, the vein of Galen, the internal cerebral vein, and the posterior cerebral artery in the posterior incisural space **(e, f)**.

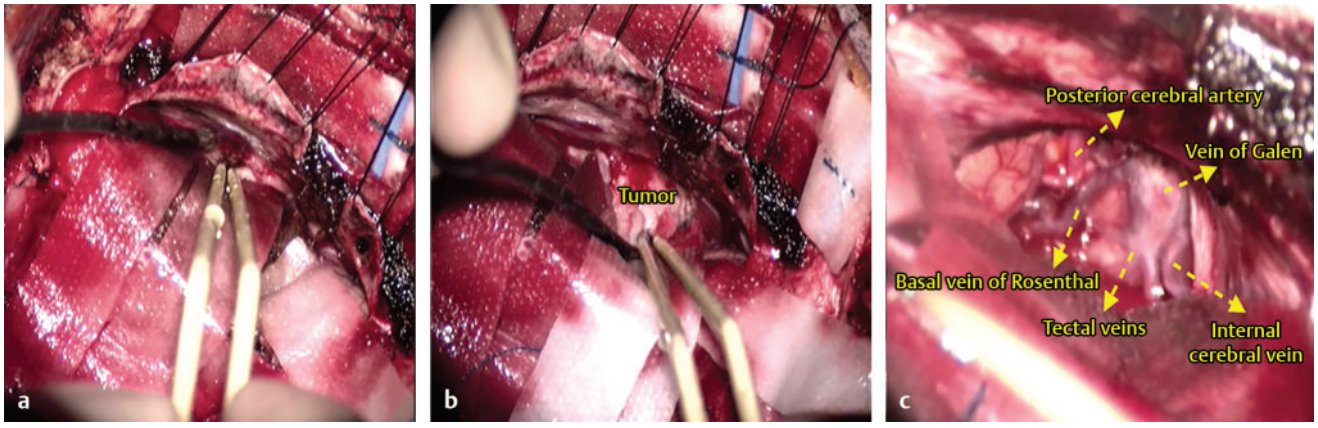


Fig. 8.13 Intraoperative pictures of the case. (a) Initial intradural exposure over the left cerebellum. **(b)** Visualization of the tumor after dissecting the supracerebellar space. Also, note that multiple moist Telfa patties have been placed over the cerebellum to minimize damage. No self-retaining retractors were used through the case. **(c)** View of the vasculature after tumor removal.

■ Description of the Technique

Electrodes for motor, somatosensorial, and brainstem auditory evoked potentials were inserted. The patient was then placed in the prone position with flexion of the chin. A midline suboccipital incision was made from 2 cm above the inion down to C4 level. We tailored the craniotomy to be more eccentric to the left side, since most of the tumor was located on the left. It is important to note that exposing the transverse sinus with the craniotomy was essential for retraction of the dura for wide exposure in this approach.

After opening the dura, tack-up sutures were placed to hold back the dura to provide access to the base of transverse sinus (**Fig. 8.13**). The dura was explicitly not opened up over the cerebellum so that the cerebellum would not be strangulated by the cut edge of the dura. The arachnoid was dissected over the superior aspect of the cerebellum without using self-retaining retractors, and the tumor came into view (**Fig. 8.13b**). The tumor appeared to be calcified and quite hard. Circumferential dissection and internal debulking were then performed to reach the tentorial attachment of the tumor. Because the tumor was adherent to the vein of Galen, this required careful microsurgical dissection under high magnification using the operating microscope. After the resection of the infratentorial portion of the tumor, it was necessary to incise the tentorium on the left side. Large pieces of the tentorium along with the supratentorial portion of the tumor were then removed. During such resections of the tentorium, it is critical to note the locations of the trochlear nerve and the basal vein of Rosenthal in order to avoid damage to these structures. Throughout the procedure, the somatosensorial, motor, and brainstem auditory evoked potential remained unchanged.

■ Aftercare

The patient tolerated the procedure well and her postoperative course was uneventful. She was discharged home soon after surgery. Pathology revealed a WHO grade I meningioma. Postoperative MRI showed total resection of the tumor with no evidence of recurrence over a subsequent 10-year period (**Fig. 8.14**).

■ Potential Complications and Associated Management

The SCIT approach is well-suited for midline tumors of the PIS, pineal region, and tectal plate. If the tumor extends more

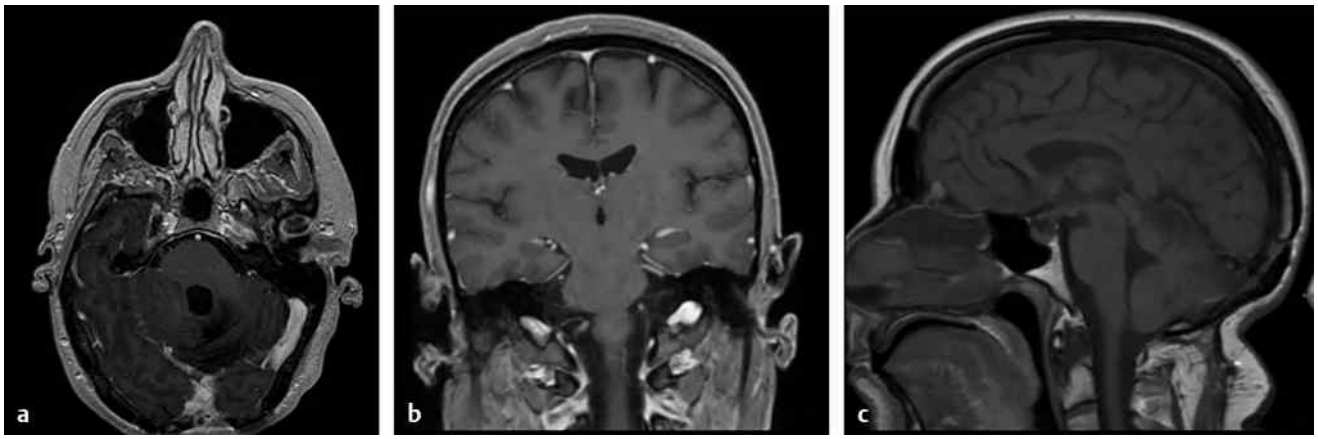


Fig. 8.14 Postoperative images. (a) T1 postcontrast axial and **(b)** coronal MRIs showed gross total resection. **(c)** T1 sagittal MRI showed that the mass effect on the midbrain and cerebellum was much improved.

superiorly or laterally above the tentorium, reaching this portion of the tumor via this approach might be difficult. Some of the tumors of the PIS, such as pineal cell tumors, can be highly vascularized. To prevent hemorrhagic complications, meticulous hemostasis is crucial. Care should also be taken to avoid injury to the transverse sinus and torcula during the craniotomy phase, and to the vein of Galen during the tumor removal phase. In case of a small tear, gentle compression with hemostatic agents like Gelfoam or surgical hemostatic agent should be applied. In the event of a large tear, the surgeon should consider microsuture repair or reconstruction. If the sitting position is utilized, it is crucial to take measures to prevent air embolisms or to treat air embolisms should they occur.

One of the most challenging aspect of tumor resection from the PIS is resecting the inferior portion adjacent or adherent to the dorsal midbrain. Dissection here may require downward pressure on the cerebellum, which has the potential to cause contusions in the superior aspect of the vermis. Thus, the use of self-retaining retractors should be avoided or minimalized.

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9 Mesial Temporal Lobe

Garni Barkhoudarian and Daniel F. Kelly

Keywords: mesial temporal lobe, ambient cistern, tentorium, endoscope, sitting position

■ Case Presentation

This patient was a 71-year-old man who had a history of primary scalp melanoma which was resected 3 years prior to our initial evaluation (T1aN0M0). He has a past medical history of diabetes mellitus (type 2), antiphospholipid syndrome, and coronary artery disease with coronary stent placement 11 years previously. No adjuvant therapy was deemed necessary at that time. Three years later, he developed a chest wall mass that was biopsied and found to be consistent with metastatic melanoma. A thorough work-up at our cancer center identified multiple metastatic lesions including the chest wall lesion, a mesenteric lesion, multiple lymph nodes, and multiple subcutaneous lesions. In this work-up, he complained of intermittent headaches and left-sided visual disturbances. On neurological examination, he demonstrated a left superior quadrantanopia (left "pie-in-the-sky" visual field defect). Hence, a magnetic resonance imaging (MRI) of the brain was obtained with and without gadolinium enhancement (**Fig. 9.1**).

■ Diagnosis and Assessment

The patient's MRI demonstrated a solitary 2.5-cm right medial temporo-occipital–enhancing lesion. T1 sequences were hyperintense suggesting either hemorrhage or melanin content. Gradient-resonance echo imaging demonstrated regions of hypoattenuation, further suggesting a hemorrhagic component. There was vasogenic edema on T2, and postcontrast sequences showed enhancement of this lesion (**Fig. 9.1**). Diffusion tensor imaging (DTI) tractography demonstrated the optic radiations just lateral to the lesion (**Fig. 9.2**). Differential diagnosis included metastatic tumor (likely metastatic melanoma), glioblastoma multiforme, recently hemorrhagic cavernous malformation, and spontaneous hemorrhage in evolution.

Given the patient's history of recently diagnosed metastatic melanoma, the high incidence of intracranial metastases in melanoma patients (> 40%), and the imaging characteristics, this lesion was thought to be consistent with metastatic melanoma. The patient had favorable markers that suggested potential response to anti-PD1 therapy (pembrolizumab). As the intracranial tumor was solitary in nature, symptomatic, and the patient had a good prognosis from an oncological standpoint, surgical resection followed by stereotactic radiosurgery (SRS) was indicated and recommended to the patient.

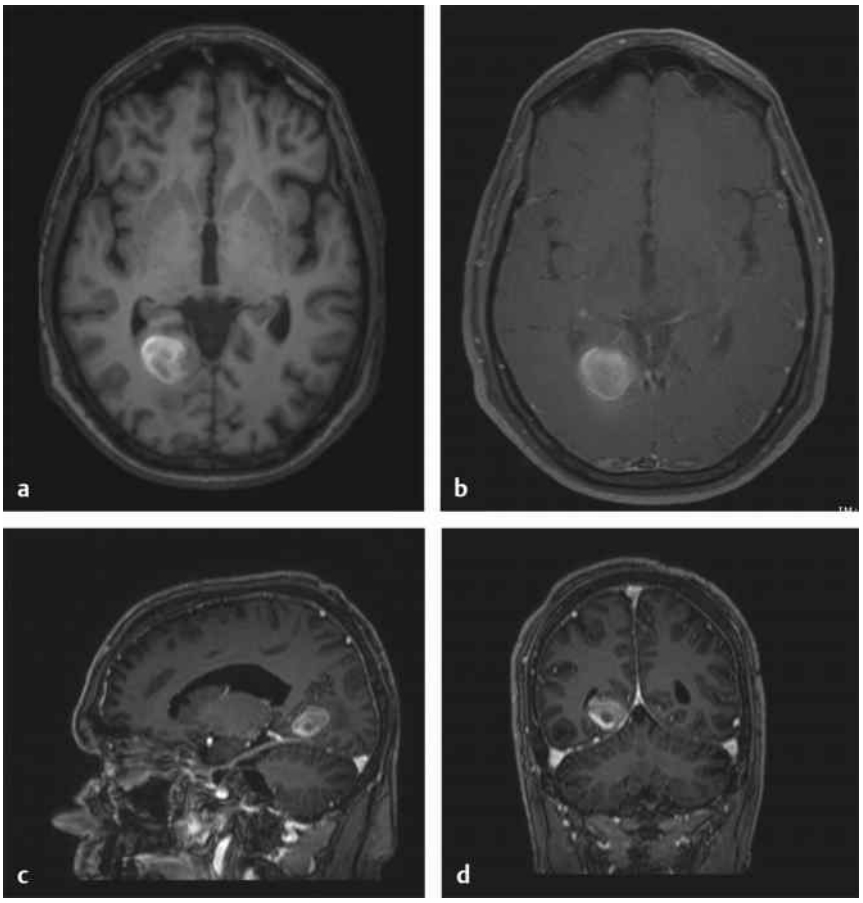


Fig. 9.1 Preoperative MRI. (a) T1 precontrast **(b–d)** and postcontrast images demonstrating a T1 hyperintense lesion in the right medial occipital lobe with minimal contrast enhancement. The T1 signal reflects both hemorrhage and melanocytes within the tumor. The tumor touches the tentorium, and its long axis is pointed medially and downward.

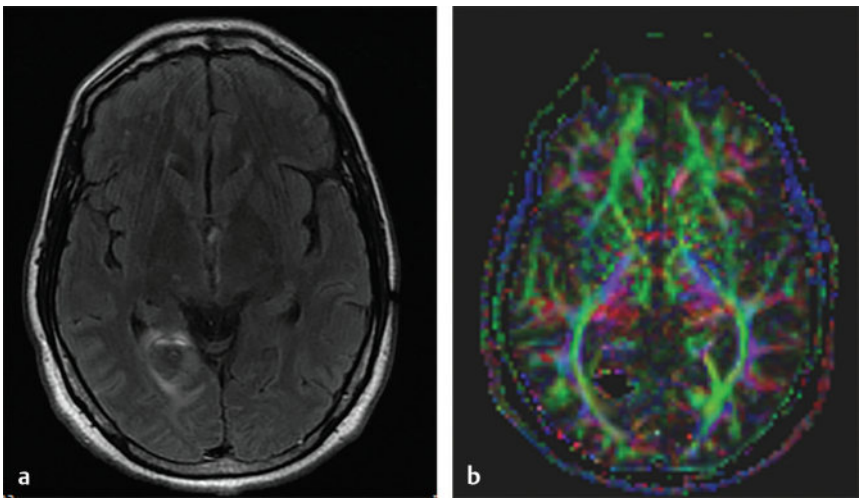


Fig. 9.2 Preoperative MRI. **(a)** FLAIR sequence and **(b)** tractography sequence demonstrating mild vasogenic cerebral edema surrounding the tumor. The optic radiations are just lateral to the tumor.

Questions

1. What was the best treatment option for this patient's metastatic brain tumor?
2. If surgery is considered, what should be the goals of surgical resection?
3. What postoperative treatment options may be helpful?

Options for Approach

The tumor was situated in the medial temporo-occipital lobe, medial to the optic radiations, which were the primary neurological structures of concern. There are numerous approach options to reach medial temporo-occipital tumors: right temporal transsulcal or transcortical approaches (through occipital horn of ventricle), right occipital interhemispheric approach, left occipital transfalcine approach, or supracerebellar transtentorial (SCTT) approach (**Fig. 9.3**). The transcortical and transsulcal operations are simpler operations, but they are also most "invasive" from a neurological point of view. These operations will almost certainly result in worsened visual field defect, possibly with a dense homonymous hemianopsia. The interhemispheric ipsilateral technique utilizes the natural plane between the falx and the occipital lobe for approach, but would require significant retraction of the eloquent cortex to reach the target. The line-of-sight to the tumor cavity would still be partially obstructed. The contralateral interhemispheric transfalcine approach offers a better line-of-sight to the target and eliminates the necessity of retraction of the ipsilateral primary visual cortex. However, this puts the contralateral occipital lobe at risk for possible retraction injury, and in this patient, the contralateral occipital lobe was the healthier one of the two.

Approach of Choice

The SCTT approach (**Fig. 9.3, Fig. 9.4**) was the ideal option in this patient given that the base of the tumor was along the tentorium and the angle of the long axis was medial and downward (**Fig. 9.1c, d**). This operation can be done in the prone position, but would require some degree of cerebellar retraction and increase the risk of retraction injury or possible stroke. Alternatively, the operation can be performed in the sitting position, allowing passive, gravity-assisted displacement of the cerebellum. The risk of venous air embolism associated with this position has been well-documented, but with vigilance from both the surgical and anesthesia teams, this risk can be mitigated.

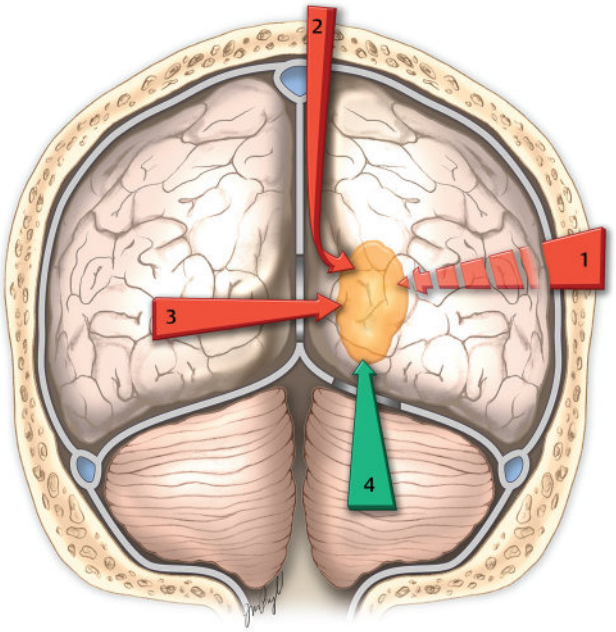


Fig. 9.3 Surgical options. 1, Right transcortical approach (through the occipital horn of the ventricle); 2, right occipital interhemispheric approach; 3, left transfalcine approach; 4, the authors' preference, supracerebellar transtentorial approach.

The patient underwent the standard preoperative medical and cardiac evaluations. Given his coronary artery disease, he was evaluated and found to have no evidence of inducible cardiac ischemia. His stents were placed 11 years ago, and it was thought to be relatively safe to stop antiplatelet medications for the operation. He underwent a routine cardiac echocardiogram

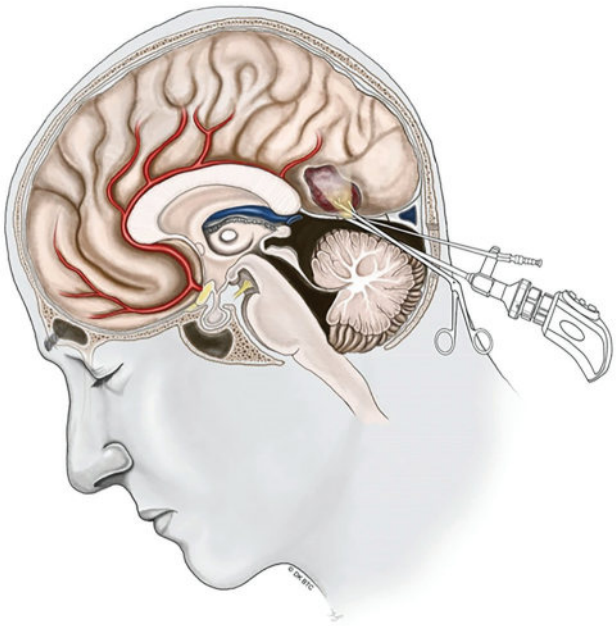


Fig. 9.4 Illustration depicting the endoscope-assisted supracerebellar transtentorial approach. (Used with permission from DFK Neurosurgical Inc.)

with bubble study which ruled out right-to-left atrial shunting, a contraindication for the sitting position.

Questions

1. What are the pertinent anatomical structures to consider?
2. What surgical approaches can help minimize injury to these structures?
3. What potential complications could occur with these approaches?

■ Description of the Technique

Position

This operation is equally defined by the position as well as the surgical technology. The sitting approach requires a comprehensive team working in concert to ensure a safe intraoperative course and good postoperative outcome. Most critical is to have an experienced neuroanesthesiologist comfortable with the sitting position and management of venous air embolism. Secondarily, an experienced surgical assistant comfortable in neuroendoscopy is very helpful. At our institution, all sitting operations require radial artery catheter for blood pressure monitoring and arterial blood assessment. Large-bore central venous catheter with the tip at the superior vena cava (SVC) and ability for rapid venous aspiration is routinely used. Transesophageal continuous echocardiography is the preferred method for early venous air embolism detection (as opposed to precordial Doppler). Neuromonitoring is also employed for somatosensory evoked potentials and electroencephalography assessment. Accurate neuronavigation is helpful to plan for the craniotomy and the subsequent tentorial durotomy.

The Three Approach Elements

Corridor: supracerebellar, infratentorial

Craniotomy: suboccipital

Modifier: incision of the tentorium

The position of the patient requires an over-the-patient adaptor for the head clamp for three-point fixation. The back is elevated as much as possible and the head is flexed maximally (chin is kept to 3 cm from chest to prevent airway compression). The legs are slightly elevated to prevent venous pooling. The arms are padded and kept on the patient's lap. A pneumatic endoscope holder is attached to the operating table and tested to ensure appropriate degrees of freedom (see **The Three Approach Elements).**

Visualization

The start of the surgery including skin incision and craniotomy is performed under loupe magnification or with an exoscope. The dural opening and initial intradural exposure can be performed with the exoscope and/or operating microscope. The majority of the intradural surgery including tumor removal is typically performed with rigid 4-mm 0 and/or 30-degree endoscopes given the improved light delivery and surgical ergonomics. For endoscopy, we typically employ a combination of freehand technique with an experienced assistant holding the endoscope, as well as use of the pneumatic arm for endoscope positioning.

Craniotomy and Tumor Removal

The craniotomy is planned to be 1.5 to 2 cm in lateral dimension, ipsilateral to the lesion, and limited by the transverse and occipital sinuses. It is not necessary to transect the occipital sinus for this operation. In this operation, the "hockey stick"

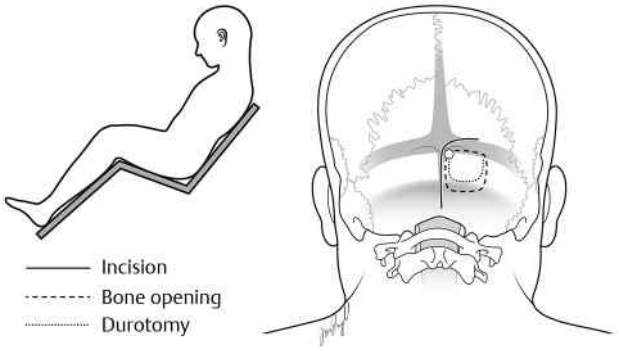
Operative Setup

Position: sitting

Incision: "hockey stick" with long limb midline, short limb to right

Bone Opening: right suboccipital, at and below transverse sinus

Durotomy: "U-shaped" based on transverse sinus



incision was used to provide adequate lateral exposure. Once the incision was created and the underlying muscles dissected, the craniotomy was performed with the burr hole at the edge of the confluence of sinuses (torcula). The Doppler ultrasound probe was used to identify the venous structures and to verify the accuracy of the stereotactic neuronavigation. Approximately 2 mm of the inferior aspect of the transverse sinus was exposed, allowing for upward displacement with the surgical instruments (**Fig. 9.5a**). Diamond drill bits were used to trim the bone edges, protecting the venous sinuses, and cauterizing small venous channels in the bone. Frequent application of bone wax was performed as the cancellous bony channels can be a source of venous air embolisms (see **Operative Setup**).

There are two separate dural incisions for the transtentorial approach. The first is the suboccipital dura accessing the supracerebellar region (**Fig. 9.5b**). The second is the tentorial dura just above the tumor. The suboccipital dura was incised in a "U-shaped" fashion. The dura was cut at the center of the craniotomy, avoiding a low incision to prevent cerebellar herniation and injury to the cerebellar cortex. It is imperative to open the dura widely in the lateral dimension. The incision should be taken up to the transverse sinus as safely as possible. Again, the use of the Doppler ultrasound is helpful in this process to define the edge of venous flow.

Once the plane was defined between the tentorium and the superior cerebellar surface, the latter was protected with cotton patties with nonstick coating (**Fig. 9.5c**). The bridging veins were identified. The aim was to preserve most of the veins; however, it is relatively safe to sacrifice a single small vein that would otherwise be in the line-of-sight and at risk for avulsion. It is preferable to sacrifice prior to avulsion, because, if unnoticed, the disrupted vein could increase the risk of air embolism.

The neuronavigation was utilized to identify the side of the tumor at the inferior surface of the tentorium. The location was verified with two-dimensional ultrasound. The tentorial dura was cut in a "U-shaped" flap based on the distal end of the tumor base. Hook knives were helpful to make this incision, as were bayoneted or single-shaft scissors. Cautery was utilized for dural hemostasis. Once the base of the tumor was exposed fully, the tumor was then entered with the usual two-handed microsurgical techniques incorporating blunt and sharp dissection as well as bipolar electrocautery when necessary (**Fig. 9.5f**). For the more superior portion of the tumor, a 30-degree endoscope and up-angled ring curettes and bipolars were particularly useful for tumor removal. The tumor was debulked and resected in a piecemeal fashion. Goals of surgery included maximal safe tumor removal and confirming tissue diagnosis. Hemostasis was achieved with typical techniques including bipolar electrocautery, thrombin, Gelfoam, Surgifoam, and hydrogen peroxide. The tentorial dura was not closed primarily, but the flap was returned to its original position. The posterior fossa surface dura was closed in typical fashion, with both inlay and onlay dural allograft placement. The bone flap was secured to the skull with titanium plates and screws to help reinforce the dural graft onlay.

The surgical techniques are demonstrated in the accompanying video (**Video 9.1**).

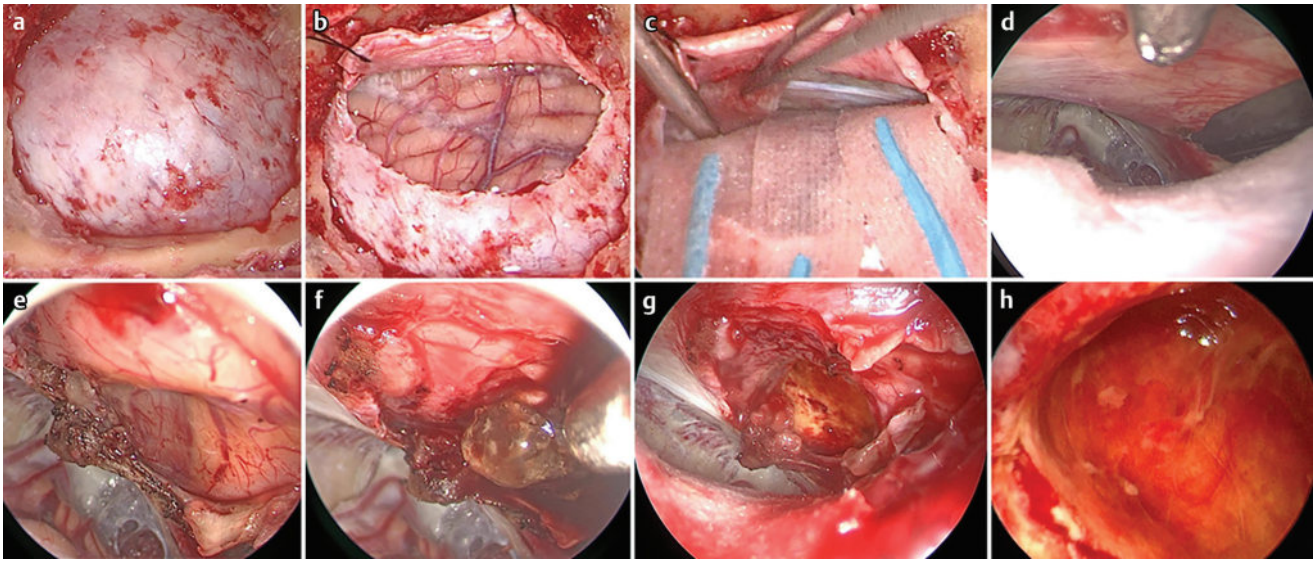


Fig. 9.5 Intraoperative images. (a) Suboccipital exposure—note the superior limit is the transverse sinus and the medial limit is the occipital sinus. **(b)** Dural flap exposing superior surface of cerebellum. **(c)** Tentorium exposed and cerebellar surface protected with cottonoids. **(d)** Tentorial location of tumor and flap development. **(e)** Medial temporo-occipital cortex exposed with tumor visible on the cortical surface. **(f)** Melanotic and hemorrhagic tumor resection. **(g)** Tumor cavity inspection (45-degree endoscope). **(h)** Tumor cavity inspection (90-degree endoscope).

Surgical Pearls

1. The primary reason to select this more complex approach (rather than a lateral transcortical or transsulcal approach) is to preserve the optic radiations and primary visual cortex on the side of the tumor. To help achieve this, preoperative identification of these structures, using anatomical landmarks and fiber tractography is performed. These data are incorporated into the neuronavigation system, which helps minimize the risk of injury and subsequent vision loss (homonymous hemianopsia).
2. Typically, the craniotomy is planned on the ipsilateral side of the tumor to minimize the downward retraction of the cerebellum, though a contralateral approach is sometimes necessary to be more parallel with the long axis of the tumor. The incision is then placed based on the position of the planned craniotomy, and either a "hockey stick" or a parasagittal incision can be used. The "hockey stick" allows for more adjustment in the lateral axis should that be necessary. However, there is a higher incidence of postoperative occipital neuralgia if the nerve is transected. The paramedian incision allows for better adjustment in the craniocaudal axis and has less chance of injuring the occipital nerve.
3. Because of the risk of air embolism associated with the sitting position, planning and vigilance are key to minimize complications. Preoperative assessment of right-to-left venous shunting should be done routinely, and both the anesthesia and surgical teams should be hyperalert during the operation.
4. Proper positioning is also key for success as there are a lot of personnel and equipment involved in this operation. Particularly, the interplay between endoscope holder, neuronavigation reference arm, and microscope must be carefully planned ahead of time.
5. The use of angled instruments (endoscope, dissectors, bipolars) are critical to success and especially important around eloquent cortex.

■ Aftercare

Unlike the majority of patients who do well after endoscopic SCTT surgery, our patient sustained an ST elevation myocardial infarction (STEMI) immediately postoperatively. He was treated supportively, as catheterization was not possible due to the fresh craniotomy. He recovered well from this STEMI, was treated with aspirin, statins, and antihypertensive medications, and was discharged without further inpatient events. He was seen 2 weeks following surgery, doing well with no neurological deficits and resolution of his preoperative vision deficits. An MRI after surgery showed satisfactory removal of the tumor (**Fig. 9.6**). SRS was planned for treatment of the tumor cavity to prevent local recurrence. Unfortunately, on the day of SRS, the patient sustained a second STEMI, and underwent cardiac catheterization which was then complicated by multiple cerebral infarctions. He subsequently died from these strokes and cardiac failure.

Though reasons for his postoperative STEMI events are debatable, certainly the temporal association of the operation calls into question its culpability. The patient had two risk factors for myocardial infarction (MI) including prior MI and a hypercoagulable state from both antiphospholipid syndrome, as well as metastatic cancer. Although air embolism is a major concern with the sitting position, its effects are immediate and do not result in delayed occlusive cardiovascular events.

■ Possible Complications and Associated Management

Potential complications with this approach include intracranial hemorrhage, venous air embolism, tension pneumocephalus, cerebrospinal fluid (CSF) leakage, and damage to the primary vision cortex or optic radiations. Given the smaller bony opening relative to other approaches, there is some concern for the possibility of deep hemostasis. Hence, vascular dissection is carefully performed, avoiding avulsion of sulcal arteries, which

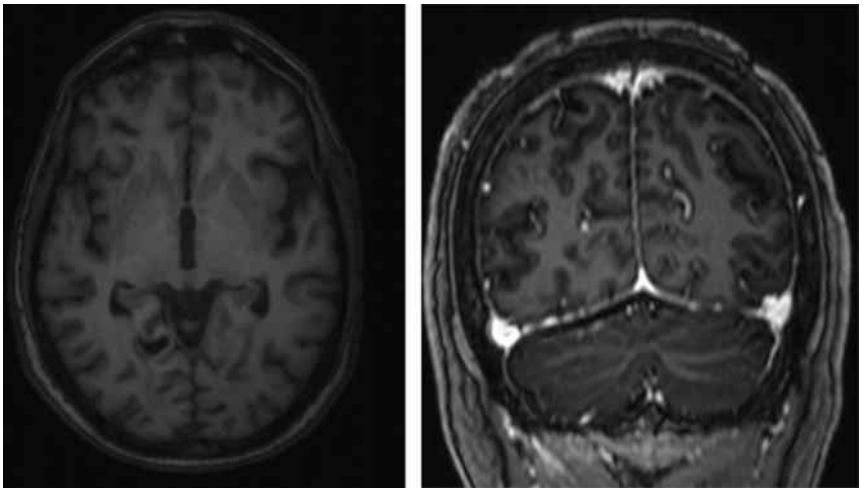


Fig. 9.6 Immediate postoperative MRI with axial and coronal sequences demonstrating gross total resection of tumor.

can retract and result in larger hemorrhages that are poorly controlled. If the occipital or temporal horn of the ventricle is entered, the CSF pathway is protected with temporary placement of collagen sponge during tumor resection and ultimately sealed with collagen sponge to prevent external hydrocephalus.

Venous air embolism is a primary concern with this operation in the seated position. As previously discussed, highly sensitive monitoring modalities (transesophageal Doppler and end-tidal CO_2 capnography) are employed, with central access to the atrium/SVC junction to aspirate air if embolism occurs. This strategy has decreased the incidence of symptomatic air embolism. Additionally, any maneuver that may result in inadvertent venous injury is minimized. The transverse sinus is only partly unroofed and is protected with the dural flap. Distal (posterior) bridging veins that would clearly be obstructing endoscope or instrument maneuverability are assessed and sacrificed to prevent delayed injury and air embolism. Larger veins are typically in midline and are protected with collagen sponge or muslin gauze.

As a routine, we obtain immediate postoperative computed tomography to identify intracranial hematoma as well as to determine the extent of pneumocephalus. Pneumocephalus is treated with 100% oxygen therapy for 24 hours. Tension pneumocephalus is a rare complication and is typically propagated with external drainage of CSF during dissection. We do not typically place lumbar or external ventricular drains for these operations. If there is obvious hydrocephalus, an endoscopic third ventriculostomy is performed (if technically feasible) prior to positioning in the seated position and the external drain that is kept in place is used for intracranial pressure (ICP) monitoring only. In the rare circumstance that the hydrocephalus is only manageable with the external ventricular drain, CSF is drained cautiously and only when required to control ICP.

The incidence of CSF leak in the absence of untreated hydrocephalus is quite low with this approach. The inlay/onlay method of closure provides adequate protection against a leak. The CSF pressures are relatively low in the region of this small craniotomy situated just beneath the transverse sinus (as compared to the craniocervical junction), which contributes to the lower CSF leak rate. At our institution, we have not encountered a CSF leak with this approach even when the ventricular system is breached.

Conclusion

In summary, the sitting endoscopic SCTT approach is appropriate for removal of tumors in the medial temporo-occipital region, particularly medial to the optic radiation. Use of angled endoscopes and instruments help reach the structures visualized, without requiring large corticectomies. Complication avoidance is achieved by meticulous preoperative evaluation and perioperative monitoring. An experienced surgical team, including neuroanesthesia, circulating and scrub nurses, as well as a seasoned surgical assistant familiar with endoscopy is necessary to perform the operation safely.

Perspective

João Paulo Almeida, Heros Almeida, Mateus Reghin-Neto, and Evandro de Oliveira

■ Introduction

The mesial temporal lobe is one of the most complex anatomical regions in the supratentorial space. Understanding of its anatomy is paramount for appropriate selection of the surgical approach and good clinical outcomes. It can be divided into three different parts for anatomical and surgical reasons (**Fig. 9.7, Fig. 9.8**). The anterior part contains the uncus and anterior part of the parahippocampal gyrus and is limited posteriorly by an imaginary line at the posterior limit of the uncus; the middle part includes the posterior part of the parahippocampal gyrus and the occipitotemporal gyrus and is separated from the posterior part by the anterior splenial line; and the posterior part is related with the basal surface of the occipital lobe and includes the cingulated gyrus, the splenium of the corpus callosum, and the inferior part of the precuneus.

Different techniques can be used for treatment of lesions located in the mesial temporal lobe region including the subtemporal, interhemispheric occipital, transcortical transtemporal, and pretemporal transsylvian approaches. However, they all have limitations. The subtemporal approach may be adequate for exposure of the middle segment and crural cistern. However, it puts the vein of Labbé at risk with the associated danger of temporal lobe infarction. This can lead to language problems if the dominant temporal lobe is involved. Transcortical approaches may be selected for approach of lesions in all the segments of the mediobasal temporal lobe region. Transtemporal resection, however, may lead to similar deficits as the subtemporal approach. Additionally, visual decline may occur if the optic radiations are injured. The pretemporal approach can expose the mesial temporal region via the transsylvian, temporopolar, and subtemporal corridors, with adequate exposure of most lesions in the anterior and middle segments. Although this approach limits the surgical manipulation of the temporal lobe, the risk of venous damage still exists, as the vein of Labbé and superficial sylvian veins may be injured when retraction is applied to expose the subtemporal and temporopolar corridors. The transsylvian corridor, with opening of the inferior insular sulcus (Yaşargil's technique) is effective for exposure of the most anterior portion of the mesial temporal lobe, but is inadequate for lesions in the posterior and basal region.

A different approach for resection of mediobasal temporal lesions is the SCTT approach. Initially described by Voigt and Yaşargil, the senior author (EO) has proposed a modification of this approach, whereby, instead of a small opening in the tentorium, a section of it is removed to extend the reach beyond the ambient cistern and mediobasal temporal lobe, to the lateral and anterior parts of the basal temporal lobe.

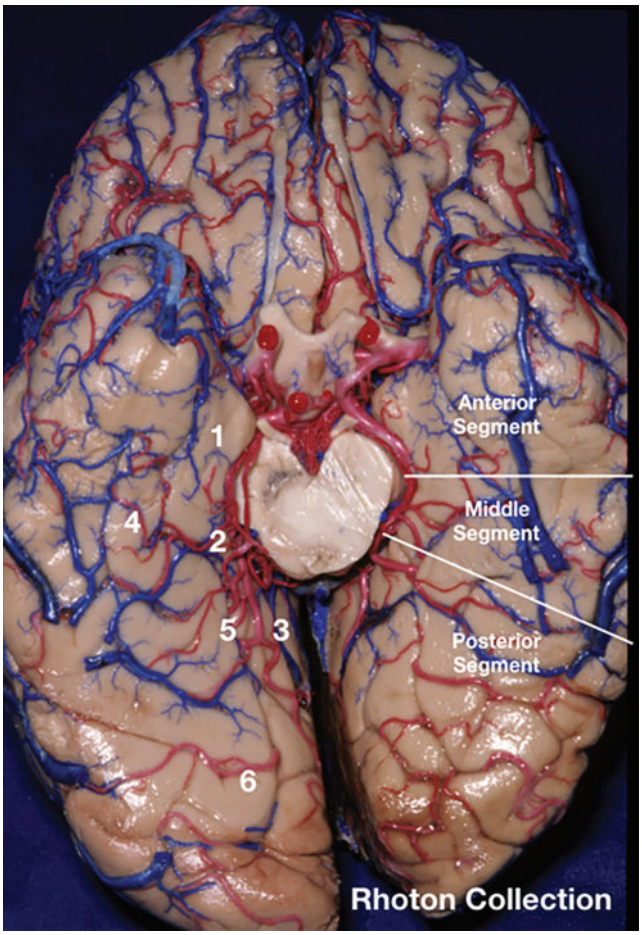


Fig. 9.7 Anatomy of the mesial temporal lobe—three segments of the temporal lobe. 1, uncus; 2, parahippocampal gyrus; 3, isthmus of cingulate gyrus; 4, occipitotemporal gyrus; 5, collateral sulcus; 6, basal occipital lobe. (The Rhoton Collection.)

■ Case Presentation

A 17-year-old adolescent girl was referred to our clinic for evaluation of a left mesial temporal arteriovenous malformation (AVM). The AVM was discovered 4 years prior, when the patient presented with intracerebral hemorrhage. She had initially undergone radiosurgery for management of that lesion, but residual AVM was discovered on follow-up, which led to the referral to our clinic for consultation.

The patient's neurological examination was unremarkable. Preoperative MRI scan and angiogram demonstrated the presence of a nidus measuring about 2 cm, seated at the posterior part of the left parahippocampal gyrus, supplied by branches of the left posterior cerebral artery (PCA) and drained by the deep venous system through the vein of Galen (**Fig. 9.9, Fig. 9.10**).

■ Anatomical and Therapeutic Considerations

The lesion was located in the posterior part of the parahippocampal gyrus. We favor the SCTT approach for resection of lesions in this location because it avoids transgression of brain parenchyma and allows exposure of the posterior part of the mediobasal temporal lobe, ambient cistern, and branches of the

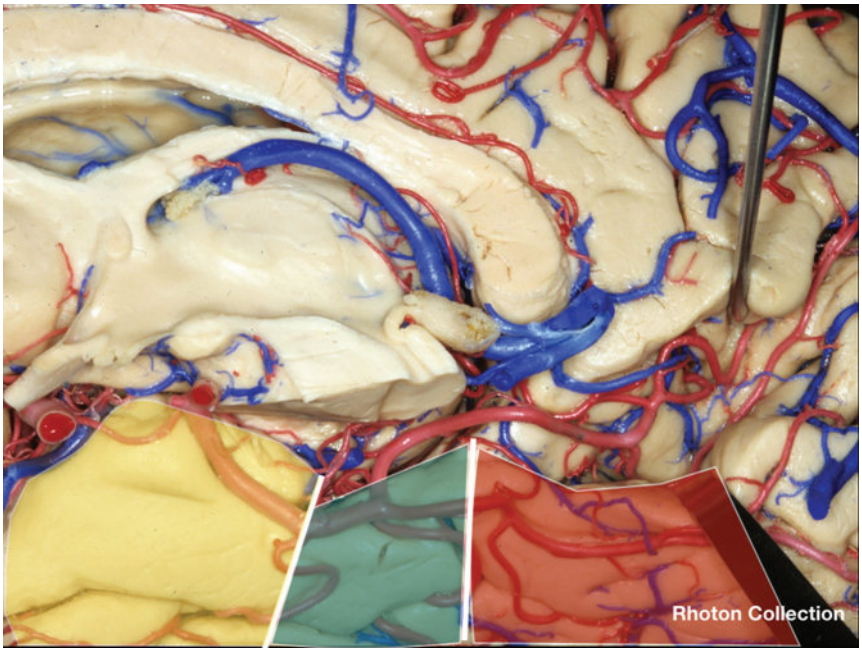


Fig. 9.8 Anatomy of the mediobasal temporal lobe (medial aspect). The anterior (yellow), middle (green), and posterior segments (red) are demonstrated. The anterior segment is composed by the uncus and anterior part of the parahippocampal gyrus, facing the anterior two-thirds of the cerebral peduncle; the middle segment is mainly composed by the posterior part of the parahippocampal gyrus; and the posterior segment is composed by the lingual gyrus, and parasplenial region, extending to the basal occipital lobe. (The Rhoton Collection.)

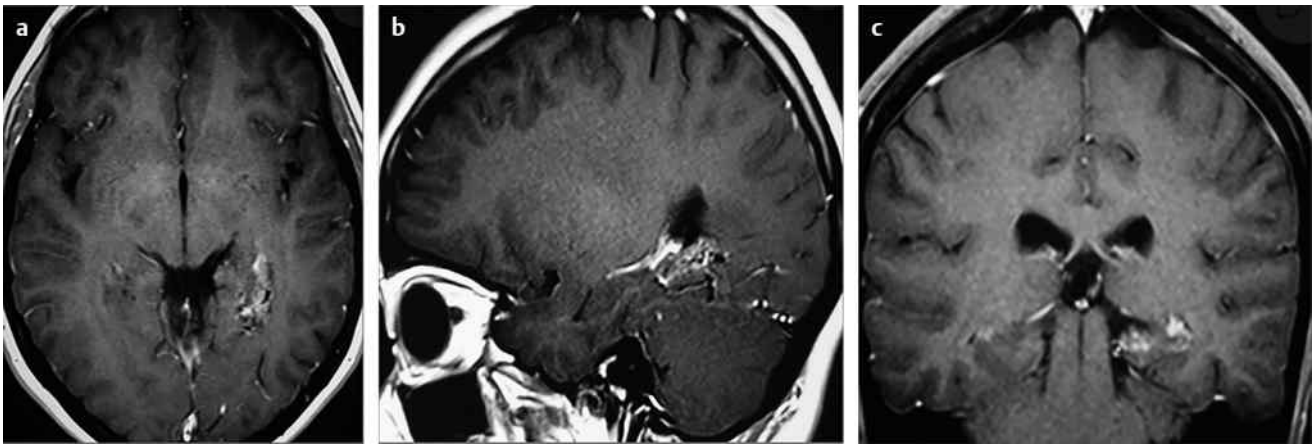


Fig. 9.9 Preoperative images. (a) Axial, **(b)** sagittal, and **(c)** coronal T1-weighted MRI with gadolinium showing an arteriovenous malformation of the left posterior mesial temporal lobe involving the posterior parahippocampal gyrus.

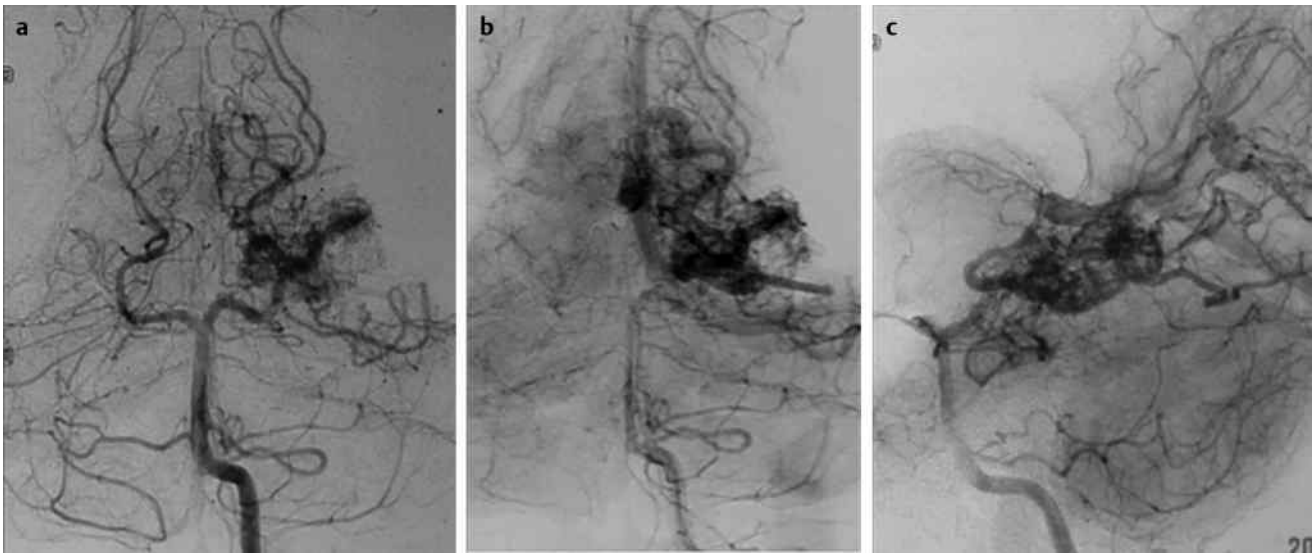


Fig. 9.10 Angiogram of the arteriovenous malformation (AVM). (a–c) This demonstrated the feeding vessels of the AVM from the left posterior cerebral artery and drainage through the deep venous system.

PCA, which supply this AVM. As previously mentioned, a wide resection of the tentorium extends the reach of the approach even further, but microsurgical experience and familiarity with the anatomy of the tentorium are critical to success.

We perform the SCTT in a semisitting position, which facilitates the exposure of the basal temporal lobe after resection of the tentorium and eliminates the need for retraction. However, it requires an expert anesthesiology team, who must be comfortable with this position and able to manage potential intraoperative complications (i.e., air embolism). For planning of surgery, besides MRI and angiogram (for vascular lesions), a preoperative echocardiogram evaluation is done in all patients to rule out the presence of a patent foramen ovale/septal defect. If that is found, we use the prone position instead.

For patients who undergo surgery in a semisitting position, central line and transesophageal echocardiogram are always used. Continuous irrigation and meticulous hemostasis throughout the procedure reduce the chances of air embolism.

■ Description of the Technique

After general anesthesia, the patient was placed in the semisitting position with the head fixed using the Sugita head holder system (Mizuho America, Inc.). The neck was flexed to expose the suboccipital region, keeping the tentorium parallel to the floor. A horseshoe skin flap was created, with the midline incision from inion to the spinous process of C2, to expose the left cerebellum (**Fig. 9.11a**). The skin and subcutaneous planes were dissected together and reflected laterally with fishhooks. The craniotomy should be wide enough to expose the transverse sinus and the confluence of sinuses to allow a retraction of the tentorium superiorly, and to expose the transverse sinus–sigmoid sinus junction (**Fig. 9.11b**).

The dura mater was opened with a U-shaped incision, exposing the suboccipital and tentorial surfaces of the cerebellum. The cisterna magna was open for drainage of CSF and relaxation of the cerebellum. One of the main advantages of the semisitting position for the SCTT approach is that it creates a natural space between the tentorium and the cerebellum with the aid of gravitational forces (**Fig. 9.11c**). Whenever possible, bridging veins should be saved and, when sacrifice is necessary, the ones from the superior aspect of the cerebellum should be divided. The veins must be coagulated as near the surface of the cerebellum as possible. Although usually not necessary, a self-retaining retractor may be placed on the culmen to increase medial and lateral exposure.

The tentorium was then resected, following the border of the transverse and sigmoid sinuses, exposing the mediobasal aspect of the temporal lobe. After exposure of the posterior part of the parahippocampal and fusiform gyri, the ambient cistern was opened for identification of the distal branches of the PCA (calcarine and parieto-occipital arteries). Those branches led to the nidus of the AVM, which was then carefully dissected. Coagulation of the feeding branches was then performed followed by coagulation of the draining veins and, finally, resection of the nidus (**Fig. 9.11d**). Careful hemostasis was achieved prior to dural closure, which was watertight to avoid postoperative CSF leak. The bone flap was replaced with use of miniscrews and miniplates, and the muscle, subcutaneous tissue, and skin were sutured in the usual fashion.

■ Aftercare

After surgery, the patient was taken to the neurological intensive care unit (ICU) and observed for 72 hours. She had no

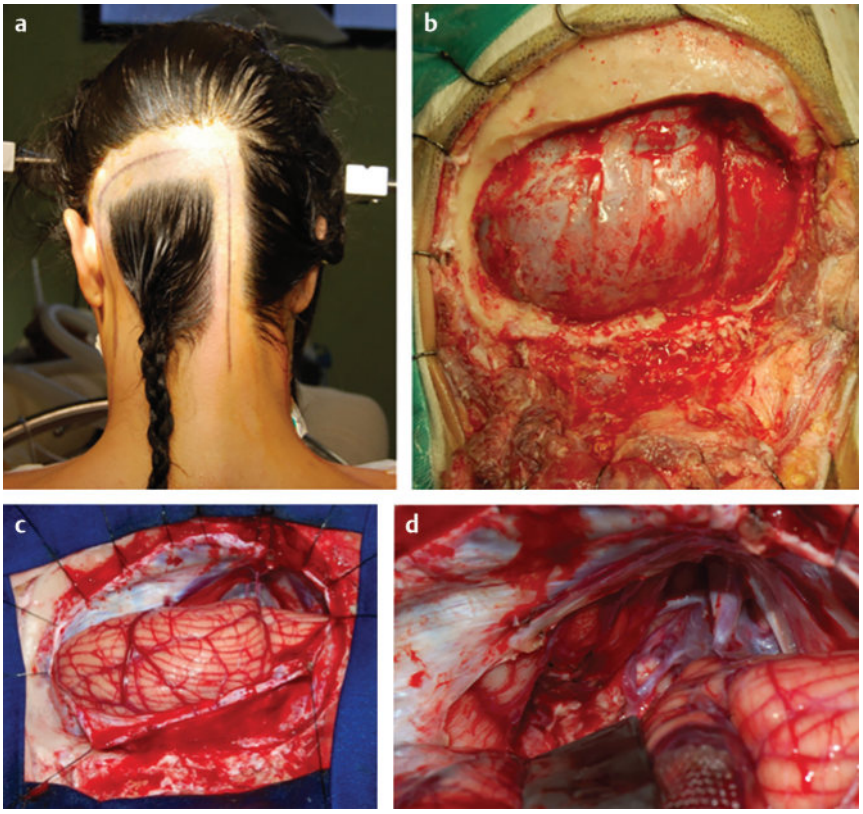


Fig. 9.11 Intraoperative image of the supracerebellar transtentorial approach.
(a) The sitting position was used and the planned incision is showed. **(b)** A left suboccipital craniotomy was performed with exposure of the torcula and left transverse sinus. **(c)** Exposure of the suboccipital and tentorial surfaces of the cerebellum after coagulation and cut of the veins in the superior part of the cerebellum. **(d)** Final view of the operative field after the removal of the arteriovenous malformation nidus. Note that a significant part of the left tentorium had been removed as well.

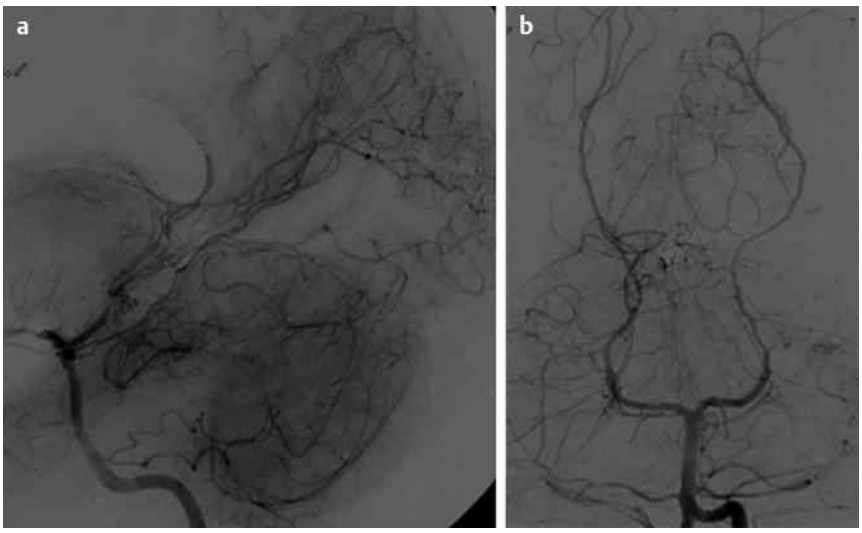


Fig. 9.12 Postoperative angiogram. (a, b) These showed that the arteriovenous malformation had been completely removed.

new neurological deficit after surgery or surgical complications. For AVM patients, we routinely observe them in the ICU for 2 to 3 days in order to maintain rigorous control of the blood pressure and reduce chances of postoperative hemorrhage. After that, the patient was transferred to the wards and underwent a postoperative angiogram, which demonstrated complete resection of the AVM (**Fig. 9.12**).

■ Commentary

In this section, we demonstrated the application of the SCTT using a vascular lesion as example, but this approach is also excellent for resection of intra-axial (low-grade and high-grade gliomas, cavernomas) and extra-axial (meningiomas, distal PCA aneurysms) lesions in this region. As discussed, we routinely use this approach for management of lesion in the posterior part of mesial temporal lobe, but it can also be used in selected cases of lesions located in the middle and anterior segments.

For lesions located in the posterior part of the mediobasal temporal lobe, besides the microsurgical approaches already mentioned, endoscopic and transtubular transcortical variants of the technique deserve to be mentioned. The endoscopic supracerebellar approach was discussed in the first section of this chapter and is an effective minimally invasive approach for resection of selected tumors in this region. In our opinion, case selection is paramount since a pure endoscopic resection should be restricted to small-to-midsize lesions that are not hypervascularized. Although endoscopy has significantly advanced in the last 10 years, hemostasis remains a challenge with endoscopic techniques and is more easily achievable under the surgical microscope. Additionally, endoscopic technique should only be performed by surgeons with sufficient experience with neuroendoscopy, since injury to major draining veins and surrounding neural structures may lead to poor outcome. That said, we believe the endoscope is a useful tool for evaluation of the surgical cavity after microsurgical resection and should be used liberally for this purpose. It may help to identify residual tumors in the corners of the surgical field and ensure total resection of tumors. Transtubular transcortical surgery, guided by neuronavigation and DTI/functional MRI may be useful in selected intra-axial cases. It is our practice, however, to avoid any transcortical approaches. Based in our experience, we recommend the use of transcisternal corridors for microsurgical resection of tumors and vascular lesions.

In conclusion, for posterior mediobasal temporal lesions, the SCTT approach is our preferred technique. If that is not possible, we favor the interhemispheric (ipsi- or contralateral) approach.

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Part IV

Tumors of the Central Skull Base

10 Dorsum Sellae *129*
11 Suprasellar *141*
12 Posterior Clinoid *151*

10 Dorsum Sellae

Jonathan A. Forbes, Charles Alex Riley, Ashutosh Kacker, and Theodore H. Schwartz

Keywords: intraventricular, craniopharyngioma, dorsum sellae, endoscopic, transsphenoidal, transcallosal, transchoroidal

■ Case Presentation

A 44-year-old left-handed man with no significant past medical history presented with headaches and memory loss, which started approximately 1 year prior to neurosurgical consultation. An elective magnetic resonance imaging (MRI) of the brain was obtained, which demonstrated a 2.4 × 2.0 × 2.0 enhancing mass perched on the dorsum sellae, extending into the third ventricle (**Fig. 10.1**). Subsequent evaluation by the neuro-ophthalmology team noted mild bitemporal inferior quadrant visual field loss. No papilledema was present.

Questions

1. What is the differential diagnosis of the MRI finding?
2. How does the differential diagnosis affect the subsequent work-up?
3. What surgical corridors can be considered for resection of this tumor?

■ Diagnosis and Assessment

The patient's MRI was reviewed in great detail with the neuroradiology team. There was no evidence of hydrocephalus. Computed tomography (CT) was performed, which revealed no associated calcifications. However, despite the lack of any cystic component or calcifications, the radiologic findings of the neoplasm were interpreted to be most consistent with a craniopharyngioma. Other pathologies (e.g., ependymoma, choroid glioma, central neurocytoma, intraventricular meningioma) were considered, but thought to be significantly less likely. The tumor was mostly solid, and entirely above the sella. The optic chiasm was anterior, and thus the tumor was completely retrochiasmal. The mass was perched on the dorsum sellae, tilting posteriorly, and involved the interpeduncular cistern. Its superior extension nearly reached the level of the foramen of Monro.

Formal endocrinology consultation was obtained and laboratory testing demonstrated evidence of secondary hypothyroidism. The patient was started on thyroid supplementation. Early evidence of diabetes insipidus was also present. The endocrinologic insufficiency strengthened the likelihood of a craniopharyngioma. An option of ventriculoscopic biopsy for histopathological diagnosis prior to definitive resection was contemplated. Ultimately, however, given the high index of suspicion for craniopharyngioma, the young age of the patient and his overall good health, the decision was made to proceed with surgery, with the goal of total resection.

■ Anatomical and Therapeutic Considerations

A variety of surgical options can be considered for resection of large tumors posited on the dorsum sellae extending in the third ventricle and interpeduncular cistern. In general, open microsurgical approaches to this region fall into two categories: under the frontal lobe through either the anterior or anterolateral corridors, or through the lateral ventricle (**Fig. 10.2**). In the former category, proceeding through the lamina terminalis would be necessary for any tumor component in the third ventricle, and in the latter, access to the lateral ventricle is obtained using either a transcortical or transcallosal corridor. When indicated, the foramen of Monro can be subsequently enlarged using transchoroidal dissection to gain wide access into the third ventricle. In rare instances in which a cavum septum pellucidum is present, a transcallosal interforniceal approach to the third ventricle can be considered. While the working depth in the transventricular approaches is considerable, and eloquent venous and neural structures are placed at some element of risk, the corridor allows for good visualization of tumor dissection from the lateral margins of the hypothalamus. In light of this versatility, the transventricular approaches are sometimes favored over the translamina terminalis approach for larger, purely intraventricular neoplasms. However, the risk of injury to the fornices, which can result in permanent memory impairment, is not insignificant.

Comparatively, the advantage of using the anterior or anterolateral corridors include better access to tumor extension

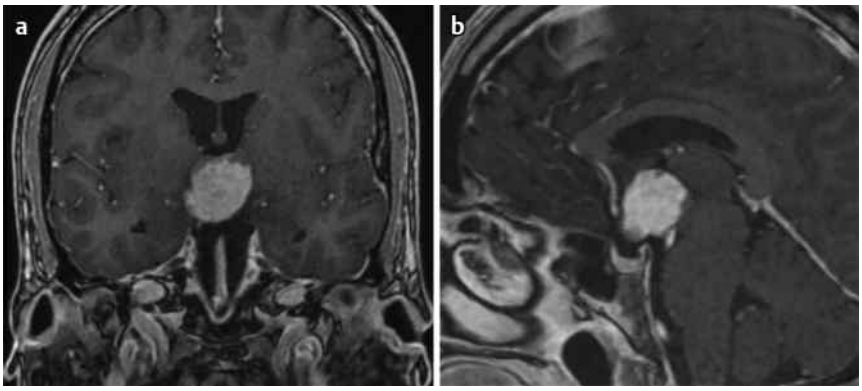

Fig. 10.1 Preoperative MRI. (a) Coronal and **(b)** sagittal T1-weighted postcontrast images of the brain which demonstrated a homogenously enhancing lesion located entirely within the confines of the third ventricle.

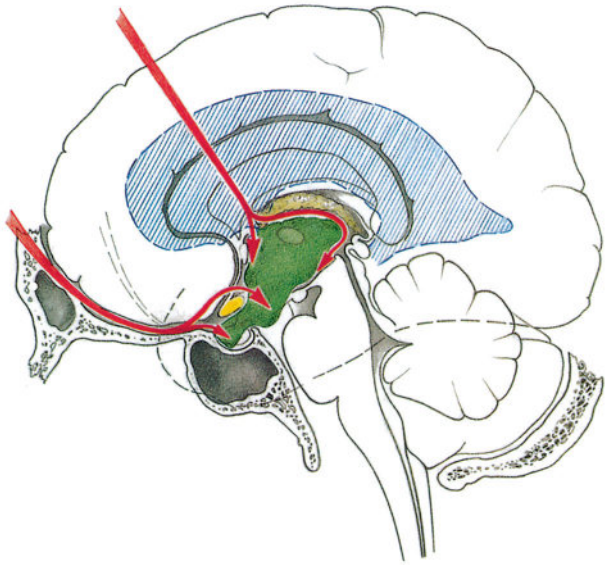

Fig. 10.2 Microsurgical approaches to the region of the dorsum sellae and third ventricle. Large tumors of this area can be approached under the frontal lobe either through the anterior or anterolateral corridors (with or without traversing the lamina terminalis). Alternatively, the area can be reached through the lateral ventricle, via either a transcortical, or transcallosal approach (with or without dissecting the choroidal fissure).

outside the confines of the third ventricle, and early visualization and protection of the circle of Willis and optic structures. However, these approaches are limited in a rostrocaudal dimension by the anterior communicating artery and optic chiasm. When approached from an anterolateral vantage, as with a pterional approach, dissection of the ipsilateral tumor margin sometimes proceeds with limited visualization. For these reasons, selection of the subfrontal approaches, with or without traversing the lamina terminalis, is sometimes limited to smaller tumors of the dorsum sellae.

Approach of Choice

Until recently, there were no reports of utilizing a purely endoscopic endonasal approach (EEA) for removing large retrochiasmal craniopharyngiomas with significant superior and posterior extensions. In fact, early publications dedicated to the advancement of expanded endoscopic endonasal methods for resection of craniopharyngiomas recommended against use of the EEA for this kind of tumor. However, in recent years, the thought on intraventricular craniopharyngiomas has significantly changed. The benefits of the EEA include superior access to ventral midline structures with minimal associated neurovascular manipulation and/or retraction. Additionally, the ability to deliver the endoscopic camera and light source to the region of tumor involvement allows for enhanced visualization of the tumor–hypothalamic interface. All these factors have translated to clinical benefits, as multiple studies have now shown superior results in patients with craniopharyngiomas treated using the EEA, especially in visual outcome. Drawbacks associated with the EEA in this setting include the learning curve associated with the use of long-shaft instruments in an endonasal setting and an increased risk of postoperative cerebrospinal fluid (CSF) fistula.

To avoid the risks of transcranial and transventricular approaches, particularly the potential damage to the fornix, the EEA was chosen for this patient.

Questions

1. What radiologic parameters should be evaluated when considering the EEA for resection of craniopharyngioma confined to the third ventricle?
2. What are the branches of the superior hypophyseal artery and their significance?
3. What corridors can be used to access the confines of the third ventricle using an endonasal approach?

■ Description of the Technique

For surgical planning, a navigation-protocol MRI of the brain with and without contrast was obtained. Review of preoperative imaging is useful for prediction of intraoperative corridors. The position of the optic chiasm was inferred using direct assessment as well as indirect approximation based on location of the anterior communicating artery. The pituitary gland was subsequently identified, allowing for calculation of the distance between the chiasm and stalk. This distance, called the infrachiasmatic interval, is commonly enlarged secondary to tumor expansion which pushes the chiasm anteriorly (**Fig. 10.3**). Also known as the chiasm–pituitary corridor (CPC), it represents the primary endoscopic surgical access to large retrochiasmal craniopharyngiomas with third ventricular extension. While important to assess preoperatively, a small CPC does not preclude removal of large intraventricular tumors using an endonasal approach. In the subset of cases associated with a narrow CPC, alternative approaches posterior to the stalk and superior to the chiasm (e.g., endonasal suprachiasmatic lamina terminalis approach) can be considered (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: transsphenoidal

Craniotomy: N/A

Modifier: traverse infrachiasmatic interval

Prior to surgery, a lumbar drain was placed and 0.25 mL of 10% fluorescein (AK-FLUOR, Akorn) was given intrathecally to help with visualization of intraoperative CSF leakage during tumor removal and again following closure. The patient was positioned supine with the head immobilized in a Mayfield clamp three-point fixation and was registered to neuronavigation. The head was elevated, tilted slightly to the left, and rotated approximately 10 to 15 degrees to the right. The nasopharynx was approached using a binostril approach. The middle turbinates were lateralized and a nasoseptal flap was harvested prior to posterior septectomy. A wide sphenoidotomy and bilateral ethmoidectomies were subsequently performed. The inferior two-thirds of the left superior turbinate was removed to create sufficient room for the endoscope. A 4 mm × 30 cm zero-degree endoscope was subsequently fixed into place in the left nostril

using a scope holder. Bony septations of the sphenoid sinus were removed using a diamond drill followed by removal of the sphenoid mucosa, which was necessary to prevent postoperative mucocele formation. The rostrum of the sphenoid was drilled down to adequately prepare for nasoseptal flap rotation at the end of the case (see **Operative Setup**).

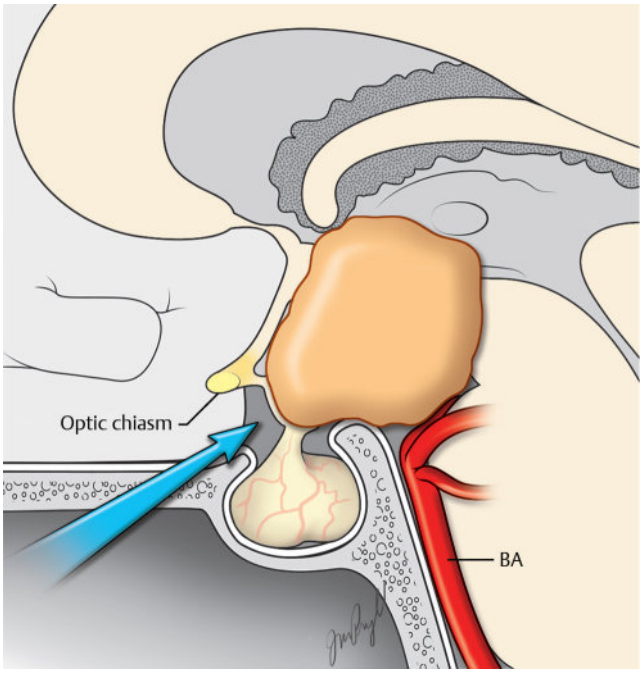

Fig. 10.3 The chiasm–pituitary corridor (CPC). This figure illustrates the surgical access between the top of the pituitary gland and the bottom of the chiasm, termed the chiasm–pituitary corridor (CPC, *blue arrow*). The CPC forms the primary surgical corridor of the endonasal approach for resection of retrochiasmatic craniopharyngioma. In one large series of patients with craniopharyngiomas who underwent endonasal resection, the mean CPC was 10.1 mm (range 5.2–19.1 mm). In this study, a small CPC did not affect the likelihood of a gross total resection. BA, basilar artery.

Operative Setup

Position: supine

Incision: endonasal approach with planned nasoseptal flap

Bone Opening: rostral sella, tuberculum sellae, posterior planum

Durotomy: dural incisions made above and below superior intercavernous sinus

Neuronavigation was used to plan the bony opening, which commonly incorporates the rostral-most portion of the sella as well as the tuberculum sellae and a small portion of the planum. This permits a more anterior–posterior oriented trajectory toward the back of the tumor. The medial opticocarotid recesses form the lateral extent of the bony opening. The bone was removed in a manner that facilitates subsequent reconstruction with "gasket-seal closure" at the end of the case (see below).

Following bony removal, the superior intercavernous sinus was isolated and cauterized with bipolar electrocautery prior to transection (**Fig. 10.4a**, **Fig. 10.5**). Residual dura was removed with Kerrison's rongeurs. The deep layer of arachnoid was then opened sharply with microscissors, revealing the underlying chiasm, stalk, and superior hypophyseal arteries and associated tributaries (**Fig. 10.4b**). Great care is taken to avoid disruption of the end-arteriole branches to the chiasmal region to prevent postoperative visual decline. On occasion, papaverine-soaked Gelfoam can be used following resection to safeguard against postmanipulative vasospasm.

Progressive tumor growth often displaces the chiasm anteriorly and widens the space between chiasm and stalk. In these cases, the interval between the chiasm anteriorly and stalk posteriorly (i.e., the CPC) forms the preferred surgical corridor. Within the CPC, it is sometimes also necessary to work in regions to the right and left of the pituitary stalk. Routinely, tumor-related expansion stretches the anatomical floor of the third ventricle to a thin translucent membrane, which is amenable to sharp dissection with microscissors. The boundary between tumor and diencephalic structures

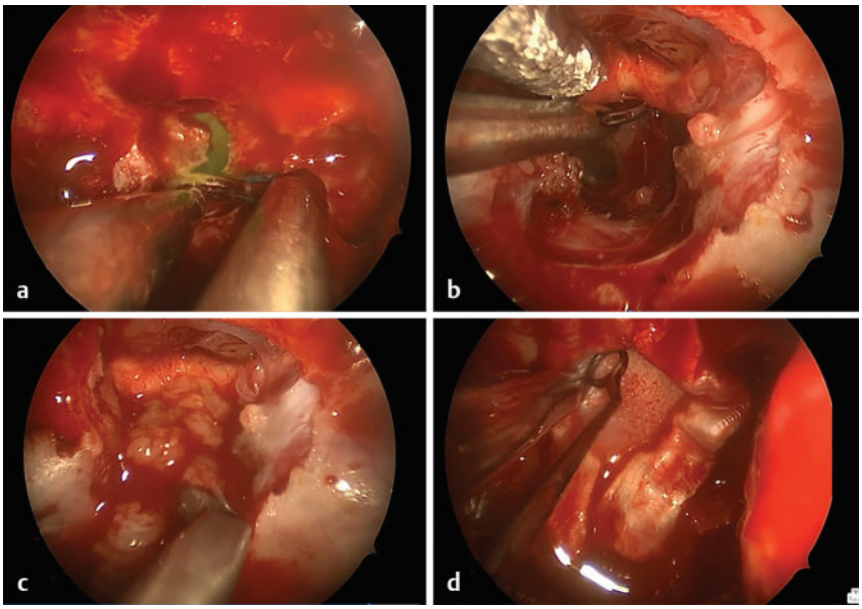

Fig. 10.4 Intraoperative views. Stepwise surgical maneuvers performed during endonasal resection of the craniopharyngioma presented in this report. **(a)** Coagulation of superior intercavernous sinus. **(b)** Sharp dissection of arachnoid inferior to the chiasm. **(c)** Piecemeal removal of the tumor. **(d)** Reconstruction using the gasket-seal technique.

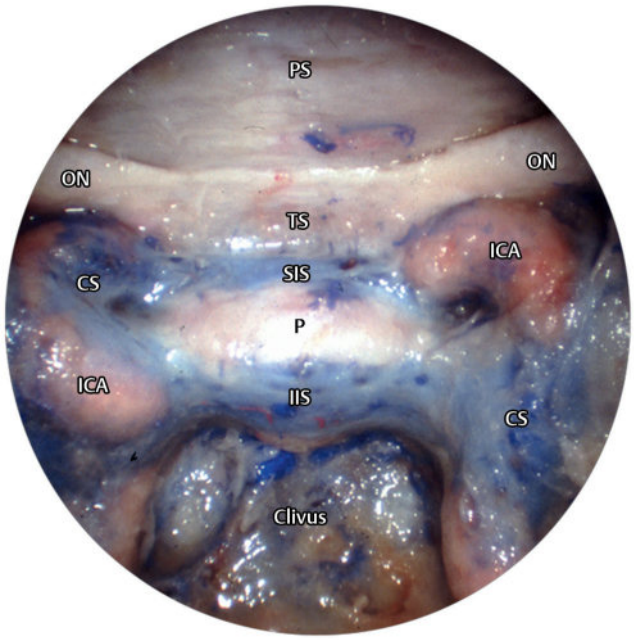

Fig. 10.5 Endoscopic view of sphenoid sinus dissection. CS, cavernous sinus; ICA, internal carotid artery; IIS, inferior intercavernous sinus; ON, dura overlying optic nerve; P, pituitary gland; PS, planum sphenoidale; SIS, superior intercavernous sinus; TS, tuberculum sellae. (Reproduced from Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery. 1st Ed. Thieme; 2011.)

is often visible during early stages of extracapsular dissection in the endonasal approach. The tumor was internally debulked (**Fig. 10.4c**) prior to additional dissection of the capsule from surrounding neurovascular structures. Direct visualization is key in safe dissection of the tumor from adjacent vessels of the posterior circulation. Angled endoscopes and instruments can be valuable to optimize visualization of the uppermost aspect of tumor extension. Every attempt was made to preserve the integrity of the pituitary stalk during tumor dissection and resection.

Following removal of the tumor, the cavity was inspected and hemostasis was ensured. The "gasket-seal" technique is utilized for closure (**Fig. 10.4d**). In this technique, tensor fascia lata obtained from the thigh was used as an onlay graft. The bony defect was subsequently reconstructed by countersinking rigid Medpor (Stryker, Englewood, NJ) in place over the fascia lata graft. The construct was covered with a nasoseptal flap, which was then buttressed with a polyethylene glycol–based hydrogel sealant. A short, edited version of operative video from this case can be found in **Video 10.1**.

Surgical Pearls

1. Neuronavigation is used to plan the bony opening, and the bone must be removed in a manner that facilitates subsequent reconstruction with "gasket-seal closure" at the end of the case. The opening must be large enough to permit surgical maneuvers, but sized and shaped properly so that it can hold the rigid Medpor graft at closure.
2. The dural opening is planned around isolation of the superior intercavernous sinus, which is subsequently cauterized and transected. As with the bony removal of part of the planum, the dural opening is planned this way to allow more surgical freedom in the anterior–posterior orientation toward the back-end of the tumor.
3. There is considerable variability of the superior hypophyseal arterial system, which joins with arterial branches from the contralateral side as well as additional branches from the posterior communicating artery to form a circuminfundibular anastomotic network (**Fig. 10.6**). In general, inspection of the superior hypophyseal artery on either side reveals individual branches to the undersurface of the chiasm, stalk, and diaphragma. Commonly, in this approach, sacrifice of the descending branch to the diaphragma is required on one side to gain sufficient access to the corridor between the chiasm and pituitary gland. All other branches should be unharmed.
4. Every attempt should be made to preserve the pituitary stalk during the tumor dissection and removal. Rarely, sacrifice of the stalk is necessary, but this is only done when the stalk has been densely infiltrated with tumor and a gross total resection is otherwise considered possible and likely. When tumor infiltration of the stalk justifies transection, this maneuver widens access to the inferior aspects of the third ventricle. If the tumor extends into the sella, the upper aspect of the pituitary gland can be removed when necessary to ensure a complete resection.

Aftercare

The patient did well following surgery. He noted a transient episode of blurry vision on the first postoperative day which resolved the next day. The lumbar drain was continued for a total of 48 hours following surgery. Postoperative MRI of the brain demonstrated gross total resection of the tumor (**Fig. 10.7**). Operative pathology returned consistent with papillary variant craniopharyngioma. The patient's anterior pituitary insufficiency worsened following surgery; he has subsequently required supplementation with hydrocortisone and human chorionic gonadotrophin (hCG) in addition to the

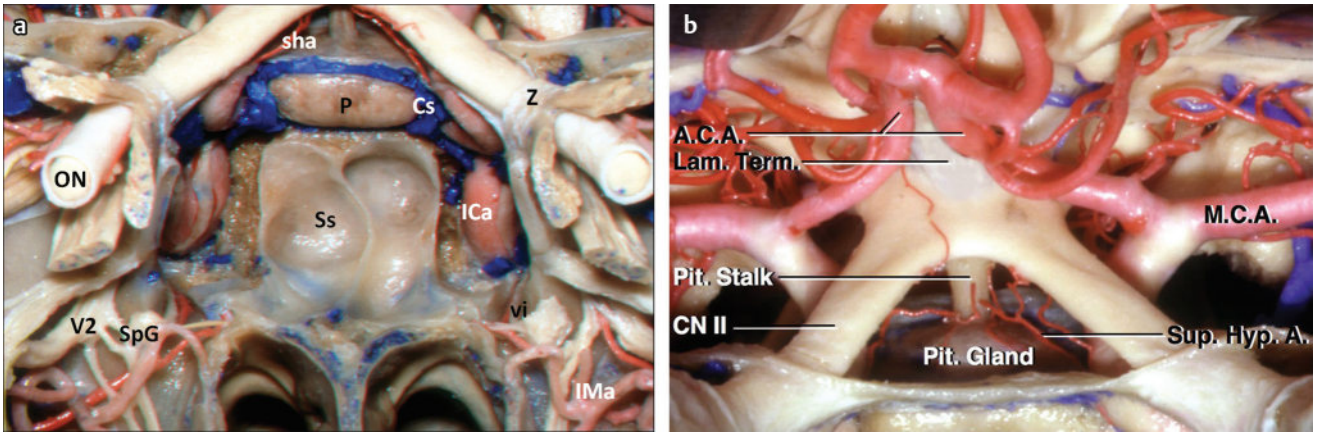

Fig. 10.6 Two views of the superior hypophyseal artery. (a) Anatomical dissection of the coronal plane. Cs, cavernous sinus; ICa, internal carotid artery; IMa, internal maxillary artery; ON, optic nerve; P, pituitary gland; sha, superior hypophyseal artery; SpG, sphenopalatine ganglion; Ss, sphenoid sinus; V2, maxillary nerve; vi, vidian nerve; Z, annulus of Zinn; **(b)** The superior hypophyseal arteries cross the chiasmatic cistern to reach the lower margin of the chiasm and pituitary stalk. ACA, anterior cerebral artery; MCA, middle cerebral artery. (Reproduced from Laws E, Sheehan J, eds. Sellar and Parasellar Tumors. Diagnosis, Treatments, and Outcomes. 1st ed. Thieme; 2011.)

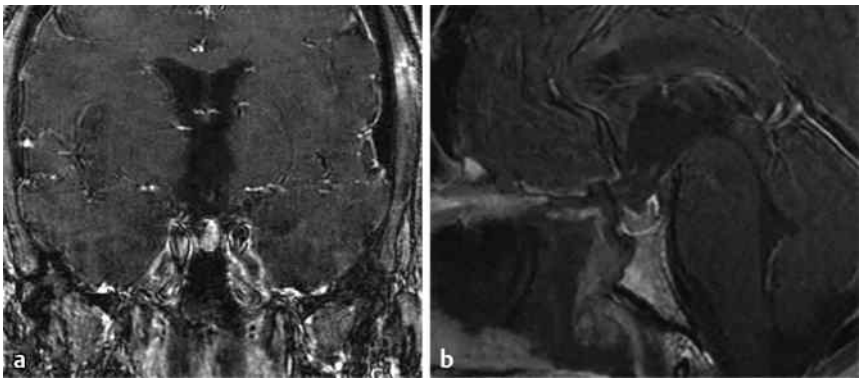

Fig. 10.7 Postoperative MRI. (a) Coronal and **(b)** sagittal T1-weighted postcontrast images of the brain which demonstrate gross total resection of the previously visualized neoplasm.

levothyroxine regimen he was on prior to surgery. To date, the patient has not developed posterior pituitary insufficiency. Follow-up neuro-ophthalmology visit 2 months after surgery demonstrated improvement in his original bilateral inferior temporal quadrantanopia.

■ Possible Complications and Associated Management

A variety of complications are sometimes encountered following expanded endonasal approaches for resection of craniopharyngiomas. Given the often intimate relationship between the tumor and the stalk, temporary or permanent deterioration of anterior and/or posterior pituitary function can occur after tumor removal. Postoperative posterior pituitary insufficiency often manifests in the form of diabetes insipidus (DI), which is characterized by an inability to appropriately concentrate urine. In a patient with DI who is otherwise alert, the high-volume loss of hypotonic fluid can be addressed in most cases with unrestricted access to free water. In the setting of florid DI, desmopressin (DDAVP) is often considered to decrease polyuria and associated requirements for fluid intake. Special attention is required in patients with postoperative DI when the thirst mechanism is compromised (e.g., a patient who is obtunded or comatose). In this setting, the inability to concentrate urine can rapidly lead to life-threatening levels of hypernatremia, and vigilant, periodic assessment of serum sodium and osmolality levels is required.

Endonasal surgery for resection of craniopharyngioma is also associated with some element of risk of anterior pituitary insufficiency. In cases where the index of suspicion for postoperative anterior pituitary insufficiency is high, patients are often routinely maintained on a glucocorticoid regimen prior to clinical reevaluation 4 to 12 weeks following surgery with the endocrinology team. This practice protects against the unexpected and potentially life-threatening development of cortisol deficiency that can present in a delayed manner following discharge. New, postoperative hypothyroidism can be assessed 3 to 7 days after surgery with a free T_4 serum measurement. Thyroid supplementation is usually initiated immediately after insufficiency is detected. Growth hormone deficiency and

hypogonadism are electively assessed in an outpatient setting by the endocrinology team, as diagnosis is not accurate in the immediate postoperative period, and the need for replacement is nonurgent.

Detection of postoperative CSF fistula is a necessary component of the routine care of patients who have undergone EEA for resection of intraventricular CPA. When intrathecal (IT) fluorescein has been administered, the characteristic fluorescent appearance on a nasal gauze dressing often makes the occurrence of early postoperative CSF leakage obvious. In cases where IT fluorescein has not been used or the leak is delayed in nature, diagnosis of postoperative CSF leakage can be more challenging and often relies on clinical evaluation. If the patient manifests drainage of clear liquid after leaning forward for a short duration of time, then CSF leak is likely. Serosanguinous drainage, on the other hand, is commonplace and is often normal. Biochemical analysis of markers such as beta-transferrin, prostaglandin D synthase, and transthyretin can be used to help differentiate CSF from postoperative nasal discharge if sufficient fluid can be obtained, but delay in attainment of these results often limits clinical applicability. In most cases, the diagnosis of postoperative CSF leakage is made using clinical findings alone.

A CT scan is often obtained when postoperative CSF fistula is suspected. Pneumocephalus is almost always present after extended endonasal transsphenoidal surgery (particularly over the anterior frontal lobes). Foci of air in the sella, parasellar, interhemispheric fissure or convexity are more suspicious but still not pathognomonic. If the nasoseptal flap is in good position, a trial of lumbar drainage is sometimes considered, as an appropriately placed flap should prevent air from entering through the defect. When this course of action is chosen, interval CT scans are recommended to safeguard against progressive increase in pneumocephalus. Prolonged CSF leakage places a patient at risk for bacterial meningitis. Because CSF rhinorrhea may not become obvious until well after discharge home, proper patient education in this regard is imperative. In rare instances, underlying hydrocephalus can predispose to multiple failed attempts at repair of postoperative CSF fistula. Ventriculoperitoneal shunt insertion may ultimately be required in this setting.

In addition to the potential complications discussed above, the EEA for resection of craniopharyngioma can rarely be associated with new postoperative neurological deficit. The new development of visual loss, cranial neuropathy, or alteration in mental status is evaluated with emergent CT of the head. When this deficit is felt to potentially relate to mass effect from hematoma or closure-related material, urgent return to the operating room for reexploration is indicated. Mild deficits that develop in a subacute manner can often be investigated using MRI.

Perspective

Walter C. Jean

■ Introduction

With the proliferation of endoscopic techniques in the recent past, multiple reports have appeared in the neurosurgical literature comparing open, transcranial surgery for craniopharyngiomas versus endonasal endoscopic operations. The majority of these reports were written by experts in endoscopy, and not surprisingly, they all conclude that endonasal endoscopic techniques are better, especially for craniopharyngiomas perched on the dorsum sellae, with large retrochiasmal components. The data cited within these reports indicate that the rate of total resection, visual outcome, and risk of postoperative seizure are all worse in the transcranial cohorts, presumably because, no matter whether the surgery is performed through the anterior, subfrontal corridor, or the anterolateral, pterional corridor, open surgery must traverse the optic chiasm and circle of Willis, and requires significant retraction of the frontal lobe to access the superior portion of the tumor. The authors also point out that with the operative microscope, illumination and visualization of the retrochiasmal component of the tumor are poor compared to endonasal endoscopy, even despite forceful retraction of brain tissue.

The main counterargument against endoscopic approaches for these deep-seated tumors is that there exists a tremendously steep learning curve to the techniques that are required to be successful. This was highlighted earlier in this chapter, and every surgical team must honestly assess its own level of comfort before tackling these highly challenging operations. The comparative reports in the literature were almost uniformly written by world-renowned endoscopy experts from ultra-high-volume centers of excellence, and that calls into question whether the results in these studies can be duplicated realistically in other practice settings. This doubt probably applies to complex craniopharyngiomas, situated entirely above the sella turcica, tilting back from the dorsum sellae, because of two main reasons. The first is that the endonasal endoscopic trajectory to the tumor go through the sella turcica, up against a presumably healthy pituitary gland, and the mobilization of the gland and stalk without damage to endocrine function, while possible, requires significant amount of experience and skill. The other reason has to do with the anatomical relationship between the tumor and the circle of Willis. The basilar artery is almost invariably draped behind the posterior capsule of the tumor and both posterior communicating arteries may be adherent to the capsule on the sides. Injuries to these vessels, especially the basilar artery, would lead to bleeding under arterial pressure that would be hard to control in the endonasal window, even in the hands of the best endoscopic skull base surgeons.

For these two reasons, my personal preference for these dorsum sellae tumors remains the use of open microsurgical techniques.

■ Case Presentation 1

A 27-year-old woman presented with a history of a craniopharyngioma which was resected twice during the year prior to presentation at our hospital. Both operations were performed via the left anterolateral corridor, and afterward, she was blind in the left eye and treated with DDAVP for diabetes insipidus. Serial MRI showed significant progression of the tumor residual (**Fig. 10.8**).

Anatomical and Therapeutic Considerations

This tumor extended from the sella turcica and dorsum sellae upward and reached beyond the level of the foramen of Monro. The midbrain and the basilar apex were pushed posteriorly, as the posterior portion of the tumor involved the interpeduncular cistern. As with any such mass lesion at the dorsum sellae, this tumor has two critical surfaces: one is its origin the suprasellar, hypothalamic region, and the other is inside the ventricle. An anterolateral approach from below to reach the origin first offers the advantage of visualizing and protecting the optic chiasm early in the procedure, but would require significant retraction on the frontal lobe to reach the superior tip of the tumor of the third ventricle. Furthermore, this corridor has been used twice before, and scar tissue may make the dissection quite challenging. This same scar tissue

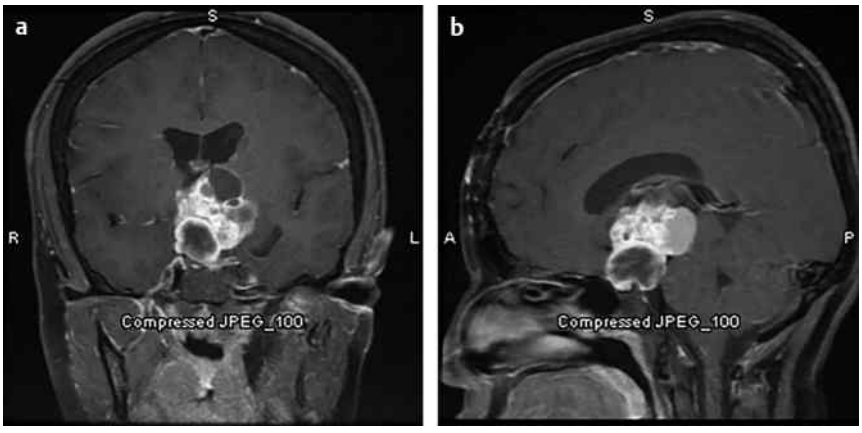

Fig. 10.8 Preoperative MRI of Case 1. **(a)** Coronal and **(b)** sagittal image showing retrochiasmal tumor extending superiorly from the dorsum sellae. This tumor had progressed dramatically over 1 year, since this patient's latest surgery.

was also a concern for coming from below endoscopically, as tumor dissection during this approach would also begin anteriorly in the area twice explored.

Alternatively, a transventricular approach would reach the superior, ventricular surface of the tumor, which in this patient was "virgin" territory. However, it has disadvantages as well, as both the optic chiasm and circle of Willis are on the far end of the tumor and thus harder to protect during the resection. Weighing all three options, it was the great height of the tumor, and the twice-used and likely ragged corridor, which ultimately determined the choice for a transventricular approach.

Description of the Technique

Later on, in this book, we will dedicate an entire chapter to the transcallosal, transchoroidal approach (see **Chapter 31**), and therefore will only describe it briefly here. The patient was placed supine and head neutral. A "U-shaped" incision was made based laterally to expose the right coronal suture from the midline to the superior temporal line. One burr hole was made on the coronal suture at the midline, and two more were done, one anterior and one posterior to this. A right frontal bone flap was elevated.

The dura was incised based medially and the interhemispheric fissure was then carefully exposed with attempts to preserve any bridging veins from cortex to superior sagittal sinus. Dissection of the interhemispheric fissure deepened until the anterior cerebral arteries were found and protected, and the corpus callosum was visualized. Traversing the corpus callosum, an opening was made into the left lateral ventricle.

The tumor was peeking through the foramen of Monro, and the choroidal fissure behind it was identified. The tenia thalamus was dissected next to open the fissure, proceeding posteriorly (**Fig. 10.9**). Once sufficient exposure to the tumor was obtained, this maneuver was halted. The cysts of the tumor were decompressed to allow exploration of the tumor posteriorly. Once the posterior margin was protected with cottonoids, the tumor was debulked and resected. The margins of the tumor were significantly adherent to both thalami on either side, which was not unexpected given the patient's history. However, with gentle and patient dissection, the whole tumor was removed. The end of the resection yielded a beautiful view of the basilar apex and left Dorello's canal (**Fig. 10.10**). A short, edited version of the video from this operation is found in **Video 10.2.**

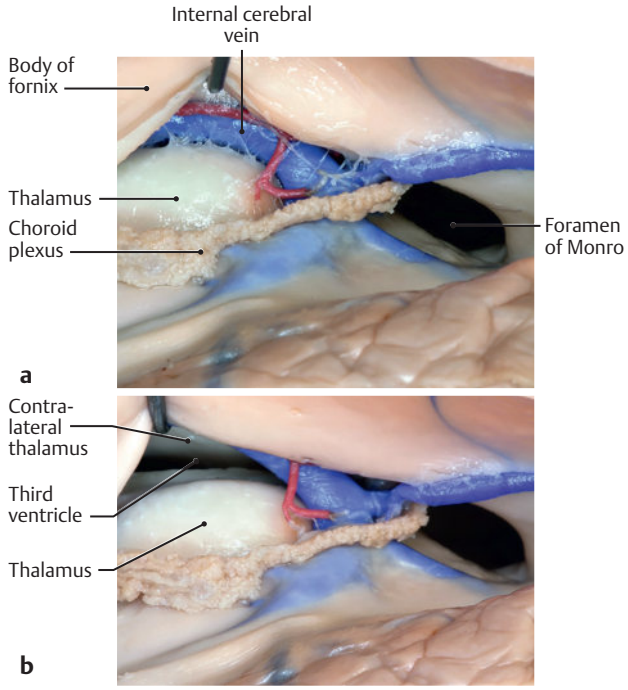

Fig. 10.9 The transchoroidal approach. (a) View of the lateral ventricle and its relation to the thalamus, fornix, and choroid plexus. The choroidal fissure can be opened between the fornix and the choroid plexus (tenia fornicis) or between the thalamus and the choroid plexus (tenia thalami). **(b)** View of a transchoroidal approach with opening of the choroid fissure on the forniceal side. Opening the fissure on the thalamic side minimizes possible injury to the fornix. The choroid plexus is used as a cushion to minimize forniceal retraction while opening the choroid fissure on the thalamic side. (Reproduced from Spetzler R, et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

Aftercare

An MRI after surgery demonstrated no tumor residual. With a long hospitalization to treat her brittle diabetes insipidus, the patient gradually returned to her neurological baseline. She did experience significant postoperative cognitive and memory difficulties.

■ Case Presentation 2

An 80-year-old woman presented with a history of progressive visual decline, urinary urgency, and progressive dementia for the last few months. On examination, she was alert and oriented, but could recall only one out of three objects at 1 minute. On formal testing, she had a bitemporal hemianopsia. Her MRI also revealed a large tumor, extending from the dorsum sellae up into the third ventricle (**Fig. 10.11**). A craniopharyngioma was again the suspected diagnosis.

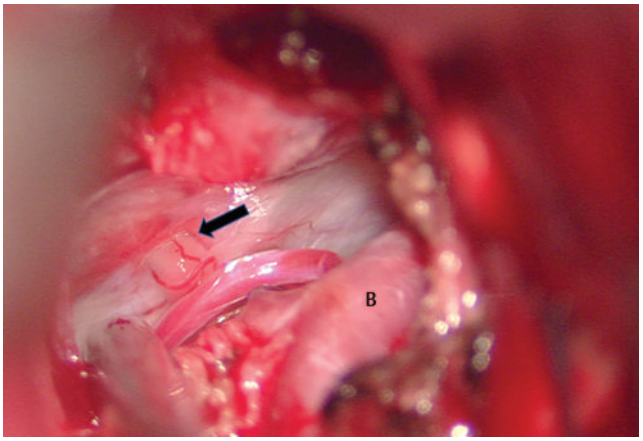

Fig. 10.10 Top-down view into the posterior fossa at the end of the resection. B, basilar artery; *black arrow* points to Dorello's canal and the abducens nerve.

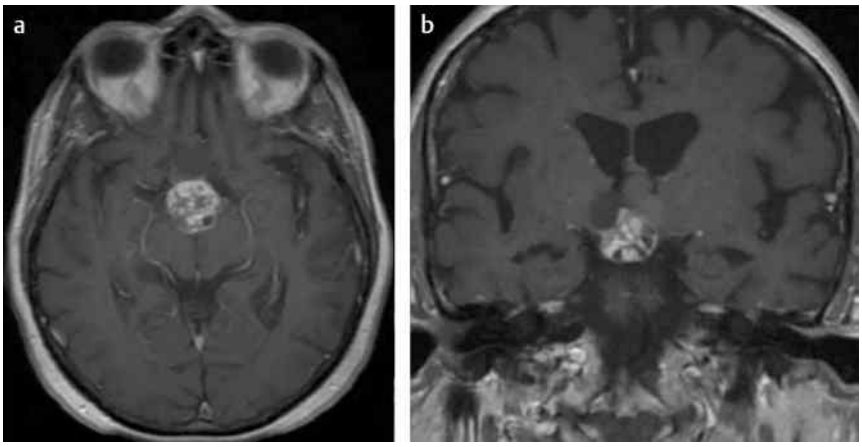

Fig. 10.11 Preoperative MRI images of Case 2. (a) Axial and **(b)** coronal images showing retrochiasmal tumor perched on the dorsum sellae consistent with a craniopharyngioma.

Anatomical and Therapeutic Considerations

At first gland, the patient's advanced age might lead one to consider symptom management over surgical therapy, but her family emphasized that she was in excellent health, and up to recently, was an independent, energetic person. The patient herself came off as an intelligent woman, clearly aware that she was losing mental clarity, and in her own way, she made it clear that she wanted to fight to hang on to her life and her vision, despite the risks, for as long as she could. As such, the plan was made for operative intervention, with the goal to at least decompress the optic chiasm to halt the decline of her vision, and to completely remove the tumor if the intraoperative anatomical conditions were favorable.

This craniopharyngioma was almost entirely retrochiasmal, behind a prefixed optic chiasm. The tumor was perched on the dorsum sellae, tipping slightly backward into the interpeduncular cistern, and the basilar apex was directly behind the tumor. In its relationship with the ventricular system, the height of the tumor reached just below the roof of the third ventricle and anteriorly, the tumor was visible in the foramen of Monro. There was no evidence of hydrocephalus.

Again, weighing the options of approach under the frontal lobe or going through the ventricle, this time there was not a clear winner like in the last example. The pros and cons of attacking this tumor from the suprasellar or ventricular surface were the same as discussed in the preceding case. The anterolateral corridor has the advantage of seeing and protecting the circle of Willis and chiasm early, but the ventricular corridor has better access to the superior portion of the tumor. However, if one combined the two approaches in a "modified above-below" technique, some of the disadvantages of each technique can be mitigated and this was done for this patient.

Description of the Technique

A small incision was made at Kocher's point on the left, and an endoscope was introduced through a burr hole and split trocar into the left lateral ventricle for visual exploration. Cystic components of the tumor were seen in the ventricle, and the endoscope was removed and replaced with the stylet of the split trocar as the rest of the operation progressed. Nearly simultaneously, on the right, a one-piece, orbitofrontal craniotomy,

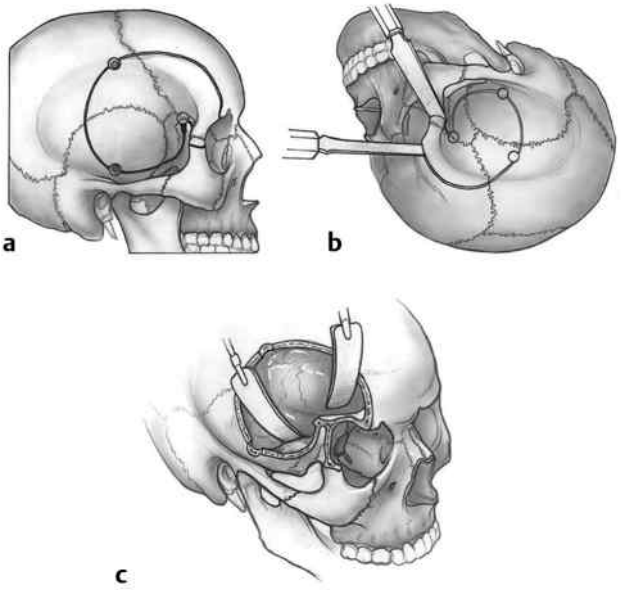

Fig. 10.12 One-piece orbitocranial opening. (a) Position of the burr holes and craniotomy. Note the extension of the keyhole at McCarty's point. **(b)** Placement of osteotomes to fracture out the flap. **(c)** Appearance of the craniotomy after bone flap removal. (Reproduced from Nader R, et al, eds. Neurosurgery Tricks of the Trade. Thieme; 2014.)

incorporating the superior orbital rim and part of the lateral orbital wall, was performed (**Fig. 10.12**). The right optic canal was unroofed by careful drilling, and once intradural, the right optic nerve was visualized and the falciform ligament above it was cut. This provided the optic nerve some added anatomical flexibility, in the hopes that manipulation of the nerve further along in the operation would be less likely to harm it with overstretching. With microscopic visualization, the initial stages of the resection were performed through the space between the optic nerves and the right opticocarotid window.

Once the resection reached the optic chiasm, the microscope was removed, and visualization of the operation was switched solely to the endoscope. It was no longer used to optimize the visualization of the tumor, as it was used in the ventricle initially, or just to gain a different visual perspective. Held in the surgeon's *nondominant* hand and placed in the anterolateral corridor, it was now used for direct visualization of the

Fig. 10.13 Surgical setup, dynamic endoscopy, and tumor resection in the principal corridor. The surgeon worked within principal anterolateral corridor, with the left, nondominant hand controlling the endoscope and the right, dominant hand working on the tumor. This provided excellent visualization of the retrochiasmal portion of the tumor.

tumor resection, and retrochiasmal resection of the tumor proceeded with a one-handed technique, as the surgeon's *dominant* hand alternated between the suction, dissector, and tumor grasper (**Fig. 10.13**). The visualization of the tumor behind the chiasm was better with the handheld endoscope than the microscope (**Fig. 10.14**).

When most of the retrochiasmal tumor was removed, the microscope replaced the endoscope in the anterolateral resection corridor again. The latter was reintroduced into the auxiliary, ventricular corridor. The light from the endoscope now served as a "homing beacon" for the microscopic resection from below, and saline pushed through the endoscope aided the removal of ventricular components of the tumor (**Fig. 10.11d**). When a dissector introduced through the principal corridor was seen by the endoscope in the ventricle, it was evident that any intervening tumor between the two corridors had been successfully removed and the operation was terminated. A ventricular drain was placed under direct visualization. Postoperative MRI revealed a complete resection of the tumor (**Fig. 10.15**).

Aftercare

The vision in the patient's right eye did become transiently worse after the operation, but it returned to baseline with brief steroid treatment. Within the year after her operation, the patient has experienced intermittent confusion and hallucinations. In

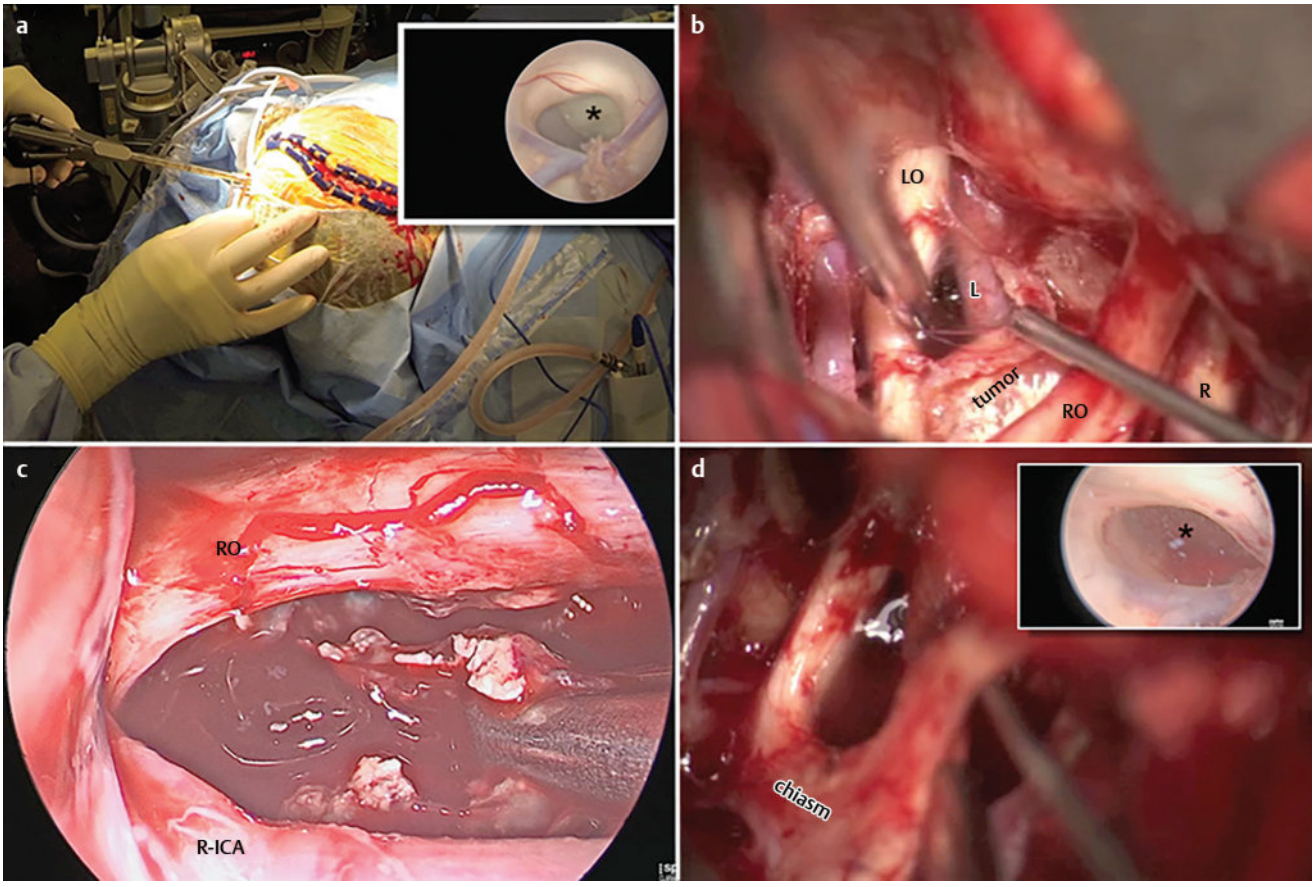

Fig. 10.14 Various intraoperative views of Case 2. (a) View of the auxiliary (ventricular) corridor via the endoscope. *, tumor cyst. **(b)** View of the principal corridor via the microscope. L, left internal carotid artery; LO, left optic nerve; R, right internal carotid artery. RO, right optic nerve; **(c)** View of the principle corridor via the endoscope, the grasper is removing tumor under the right optic nerve (RO). R-ICA, right internal carotid artery. **(d)** Combined views of the microscope of the principle and endoscope of the auxiliary corridor. *, endoscopic view of the foramen of Monro.

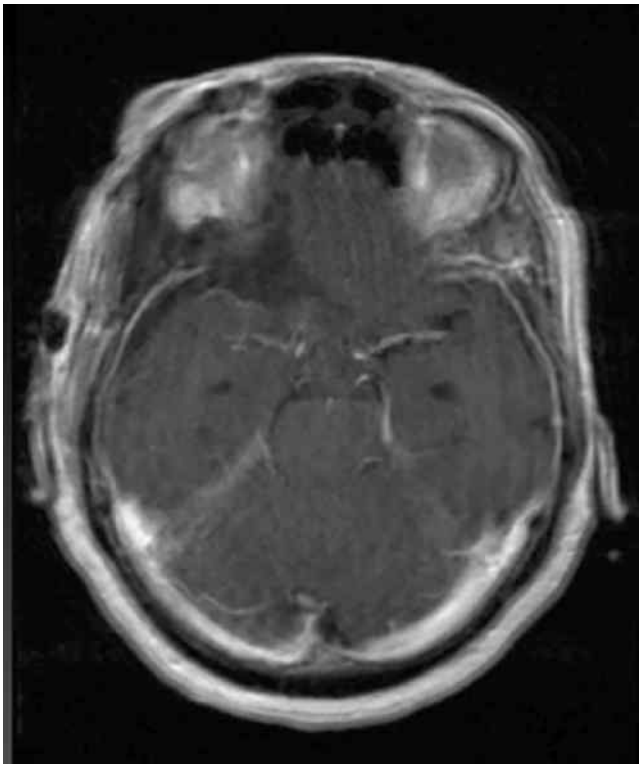

Fig. 10.15 Postoperative MRI with gadolinium. Axial view showing the tumor had been completely removed.

addition, she has had a few suspected seizures, even though electroencephalographies (EEGs) were unable to capture them. She remains on antiepileptic medications for these.

■ Commentary

The last case highlights the role that the endoscope can play in modern neurosurgery. It is not restricted to usage in the endonasal and ventricular routes, but can also be applied in "traditional" microsurgical corridors, to provide illumination, as well as visualization of the resection process. In this case example, the view behind the chiasm was far better with the endoscope compared to the microscope, and the resection under endoscopic visualization also required less retraction on the frontal lobes. Granted, holding the endoscope with the nondominant hand and switching instruments in the dominant hand to remove tumor requires some patience and experience. Furthermore, the endoscope is not designed to visualize what is behind it and once positioned to visualize the retrochiasmal space, it must be held steadily so as not to stretch the optic nerve or damage the carotid artery. Nonetheless, by combining the ventricular and anterolateral approaches, and by alternating visualization modalities, some of the disadvantages of using any single corridor alone were mitigated. Furthermore, when the two corridors were used together simultaneously, the benefits magnified even further. The light of the ventriculoscope served as a great guide for the trajectory of the microscopic dissection, and using flushes of saline (or a four-French Fogarty balloon) to push down the ventricular component, hard-to-reach portions of the tumor were "delivered" into the main resection corridor.

The recent collection of reports that compared open microsurgical and endonasal endoscopic techniques has many valid conclusions, some of which were reflected in these two case examples. In particular, they stated that transcranial operations have higher rates of postoperative seizure, poorer cranial nerve, cognitive and visual outcomes, and lengthier hospital stays. All these problems happened with our two patients. In the second case, the rare seizures took away the patient's independence, which she so cherished, and fought so courageously to maintain. Also stated in the literature reports, hypopituitarism is more likely with transcranial surgery, and this seems counterintuitive when one considers that the trajectory of endonasal approach goes right up against the pituitary gland. It would be useful to investigate how often mobilization of a previously healthy pituitary gland leads to hypofunction after endonasal surgeries. The rates of total resection and recurrence were grossly equivalent between the two major approaches, and only CSF leak rates were worse with endonasal endoscopic surgery. What few, if any, comparative reports mention is the occurrence of vascular injury. As large tumors of the dorsum sellae inevitably extend into the interpeduncular cistern and thus involve the basilar apex, protection of the basilar artery is of paramount importance. With vascular clips ready as the tumor dissection reaches its posterior extreme, there is a chance for immediate control if basilar injury occurs within a transcranial operation. The same may not be said during an endonasal operation, and a single fatal outcome may be sufficient to change a surgeon's perspective on operations in the dorsum sellae region forever.

■ Recommended Reading

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11 Suprasellar

Lai-Fung Li and Gilberto Ka-kit Leung

Keywords: pituitary adenoma, above-and-below, supraorbital keyhole, endoscopic, transsphenoidal

■ Case Presentation

A 48-year-old woman presented with the chief complaint of progressive visual loss. She has a complex history of a nonfunctioning pituitary adenoma, and had undergone multiple previous surgeries. The first surgery was a transsphenoidal (TS) operation performed 20 years prior, and it was a subtotal excision followed by external beam radiotherapy to treat the residual. She had been on thyroid hormone and cortisol replacement ever since. She underwent a second TS for recurrent disease 6 years afterward. The tumor recurred yet again 2 years later, and at that point, she underwent a resection through a transcranial interhemispheric approach which was complicated by infection and hydrocephalus. A ventriculoperitoneal (VP) shunt was placed not long after that.

Since that time, she has had two more endoscopic transnasal TS operations because the tumor regrew, showed sign of hemorrhage, and her vision deteriorated as a result. After each surgery, her vision improved despite subtotal tumor removal. After the last operation, her visual acuity on the right was 20/100, and her left eye had light perception only. Overall, she had four TS operations, one craniotomy and radiotherapy.

At the current presentation, she once again complained of visual deterioration, starting 1 month prior. Both eyes had no light perception on examination. Fundoscopic examination showed that both optic discs were pale, but fortunately, there was no sign of ophthalmoplegia. Magnetic resonance imaging (MRI) of the brain and pituitary was obtained (**Fig. 11.1a–e**).

Questions

1. Apart from tumor recurrence, what diagnoses must be considered for her visual decline?
2. Given her past medical history, what are the potential problems that would call for urgent intervention?
3. What aspects of the MRI findings impact most significantly on the decision making and therapeutic considerations at this time?

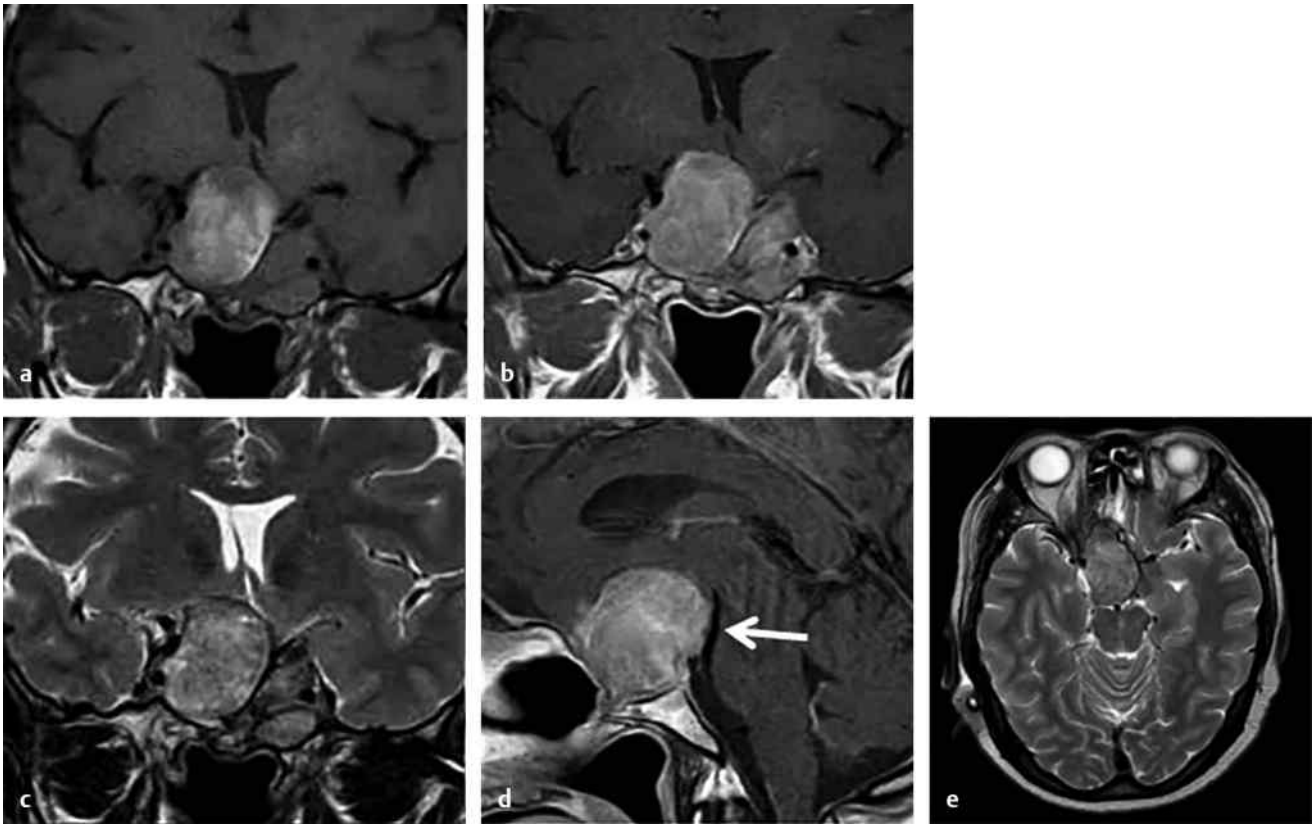


Fig. 11.1 Preoperative MRI of the pituitary tumor. **(a)** Coronal T1 sequence. **(b)** T1 sequence with gadolinium. **(c)** Coronal T2 sequence. **(d)** Sagittal T1 sequence with gadolinium contrast. Cavernous sinus involvement is better appreciated in T1 and T2 sequences prior to gadolinium administration. The sagittal view showed the basilar artery (BA) with signal void (*arrow*) in contact of posterior tumor border. **(e)** Axial T2 sequence. Posterior border of tumor abutted on BA.

■ Diagnosis and Assessment

Since the patient had a tumor with a history of repeated hemorrhages, the differential diagnoses for her vision issues would include tumor recurrence and pituitary apoplexy, both of which could produce mass effect on the optic nerves. However, given the subacute onset of visual deterioration, another important consideration is radiation-induced optic neuropathy. In patients with recurrent tumors and a complex treatment history, it is important not to bypass an endocrinological investigation. Under-replacement of her cortisol or noncompliance with medical prescription could lead to an addisonian crisis, which would require emergent intervention. Luckily for this patient, her serum electrolytes were all normal.

The MRI showed a large recurrent tumor occupying the sella and suprasellar region (**Fig. 11.1**). The tumor mass consisted of two lobules. The right lobule distorted the third ventricle, and was displacing the optic chiasm and the right A2 artery upward. Laterally, this lobule was abutting the cavernous sinus and the right internal carotid artery (ICA). Some tumor components invaded into the right cavernous sinus without passing the intercarotid line (i.e., Knosp's grade 2 lesion).

The left lobe was smaller but was nonetheless touching the optic chiasm and the left optic nerve. It encased the left cavernous ICA (i.e., Knosp's grade 4). It extended superiorly and posteriorly and displaced the basilar artery (BA) (i.e., Wilson's grade 4 and stage E). The MRI appearance of both lobules was indicative of tumor mixed with hemorrhage and more importantly, there was interval enlargement when compared with images taken 6 months prior.

The diagnosis was that of a recurrent pituitary adenoma with evidence of recent hemorrhage and optic apparatus compression. This patient has had multiple recurrences after subtotal tumor resections, and though her vision improved after every surgery, the overall trajectory of her vision was toward blindness. The option of yet another conservative subtotal resection seemed futile, as it would simply delay the inevitable. Although our primary goal was optic nerve decompression to salvage her vision, the overall surgical plan was for definitive treatment of her tumor. If any residual were to be left, it must be made small enough for radiosurgery to control it, as the patient is out of all other options.

■ Anatomical and Therapeutic Considerations

Given the aforementioned surgical goals, and our patient's complicated tumor configuration, we did not believe that a TS would achieve a radical resection because of the cavernous sinus invasion and retrosellar extension. Standard transsphenoidal surgery (TSS) is associated with a high complication rate for large/giant pituitary adenoma with suprasellar extension, because of difficulties in dissecting important structures from the tumor capsule, failure of the suprasellar tumor component to "descend" after removal of the intrasellar portion, and subsequent edema and hemorrhage of the residual tumor. These limitations could be overcome by adding on approach elements to *extend* the TS approach.

Options for Approach

The endonasal corridor can be extended in multiple directions to widen the surgical exposure (**Fig. 11.2**). Anteriorly, additional bone removal via the transplanum approach can provide better access to the anterior communicating artery and optic apparatus. Inferiorly, removal of the clivus in the transclival approach would improve exposure toward the BA. Laterally, after a medial maxillectomy and dividing the vidian nerve, a transpterygoid approach can open the lateral compartment of the cavernous sinus (**Fig. 11.3**). By combining all of these, the transsellar, transtuberculum, transclival, and transpterygoid approaches would theoretically allow complete tumor exposure.

The major challenge with our patient is her history of multiple operations, infection, radiotherapy, and repeated hemorrhage, all of which would be expected to have caused severe scaring between the tumor capsule and adjacent brain structures. Moreover, any extensive endonasal opening would require a robust repair. In our patient, not only would healing be hindered by previous irradiation, a vascularized nasoseptal flap was not possible because she had a large septal defect from previous surgery.

Alternatively, a transcranial approach would provide multiple corridors for the exposure and dissection of tumor away from critical structures. However, one major drawback of the transcranial approach is poor visualization of the intrasellar portion of the tumor. As neither a "from-below" nor a "from-above" approach would achieve safe and radical excision, we considered a combined "above-and-below" approach. This could be done in stages, which would require two separate, and likely long sessions of general anesthesia. If done in one simultaneous operation, not only would anesthesia risk be curtailed, but the two surgical teams can assist each other. The cranial surgeon can concentrate on the dissection of the tumor from vital structures and the protection of the latter against the TS surgeon's manipulations. In fact, the cranial surgeon would make every effort to avoid breaching the tumor capsule so as to reduce bleeding into the subarachnoid space, and to deliver the tumor bulk toward the sphenoid sinus. The TS surgeon would be responsible for tumor resection with guidance from above, not only via navigation but by an experienced colleague. In this scheme, the opening in the sella floor would be left intentionally limited, so that it is closable with fat packing only. At closure, the cranial

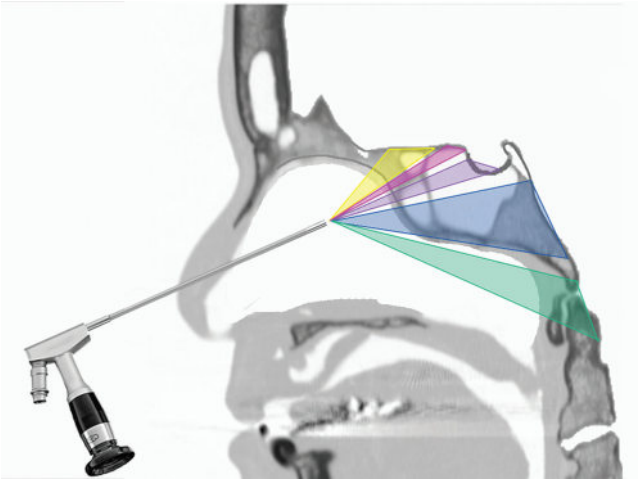


Fig. 11.2 Illustration of the anatomical structures of the anterior and middle skull base accessible through the extended endoscopic endonasal approach. *Yellow zone*, transplanum approach; *pink zone*, transtuberculum approach; *purple zone*, transsellar approach; *blue zone*, transclival approach; *green zone*, transodontoid approach.

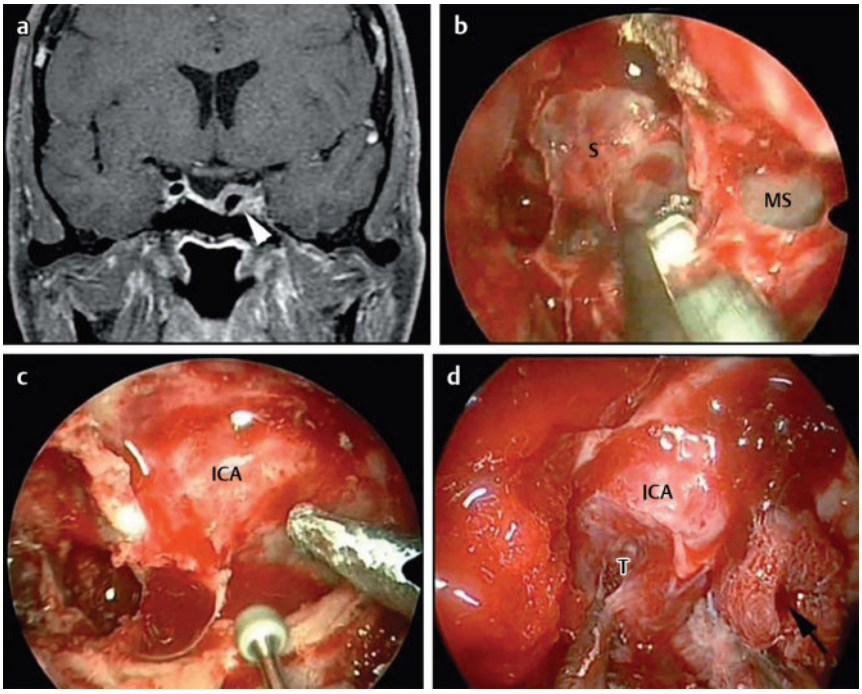


Fig. 11.3 Example of the transpterygoid approach (left). (a) Coronal MRI demonstrates a sella tumor surrounding the left internal carotid artery (*arrowhead*). Empty sella is also noted. **(b)** The posterior wall of the maxillary sinus is removed using the Kerrison rongeurs, starting at the sphenopalatine foramen. **(c)** The pterygoid process is partially removed using the drill. **(d)** Access to the cavernous sinus, both medial and lateral to the internal carotid artery, is achieved. ICA, internal carotid artery; MS, maxillary sinus; S, sella turcica; T, tumor extending posterior to ICA. (Reproduced from Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery. 1st ed. Thieme; 2011.)

surgeon can monitor the completeness of the repair, viewing from above.

Approach of Choice

With the decision made to approach "above and below" to remove the tumor, the only remaining consideration is which transcranial corridor to take. To attack tumor situated in the left cavernous sinus, our best options was an extradural Dolenc approach, which has the advantage of minimizing intradural bleeding. Given that patient's poor left-sided vision, ophthalmoplegia in this eye may not be a major concern, and if risking this means protection of the vision of the good right eye from yet another future recurrence, this risk would be well worth taking. As such, the final plan was a Dolenc approach for the left cavernous sinus tumor, simultaneous to an endonasal approach from below, with the goal to deliver the tumor bulk downward for TS removal.

Questions

1. What intraoperative neurophysiological monitoring techniques should be used if this patient has residual functional vision?
2. How would you position all the surgical personnel who must work simultaneously?
3. What are the branches of cavernous ICA that may be put at risk?

■ Description of the Technique

It is critical that the two surgical teams be positioned such that both can function properly. Otherwise, any advantage gained from a simultaneous two-team approach would be negated by hampered function of either team. The TS team stood on patient's right side and the cranial surgeon was seated at the

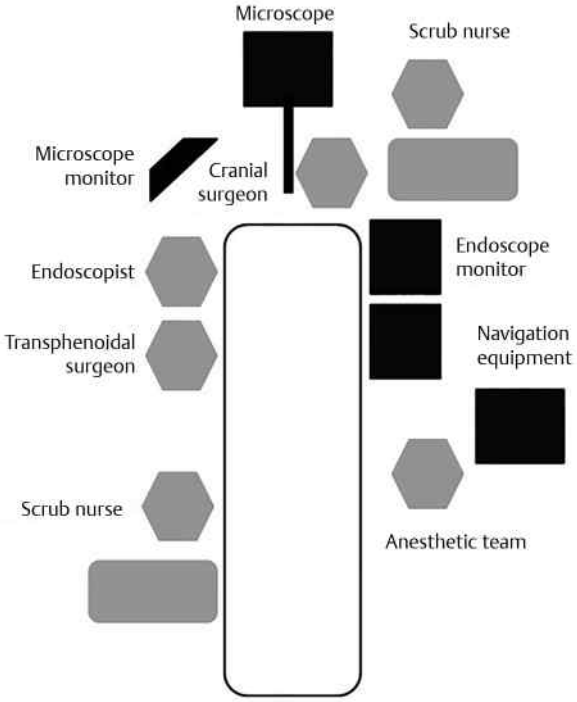


Fig. 11.4 A schematic diagram illustrating the arrangement of personnel in the operating room.

headend (**Fig. 11.4**). Both the TS surgeon and endoscopist were positioned on the patient's right and shared a monitor on the patient's left. The TS scrub nurse was also on the patient's right side. The neuronavigation computer was placed next to the endoscopic monitor, so that both monitors could be seen by both surgical teams. The cranial surgeon worked at the headend as in usual neurosurgical setting. A monitor for microscopic image was placed at cranial surgeon's right side such that it was at 45 degrees to both teams. This allowed the TS team to see the operative field of the other team. The cra-

nial scrub nurse was at the cranial surgeon's left side (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: above (anterolateral) and below (endonasal)

Craniotomy: pterional

Modifiers: anterior clinoidectomy, above-and-below simultaneous approach

Prophylactic broad-spectrum antibiotics (ceftriaxone 2 g and metronidazole 500 mg) and stress-dose cortisol (hydrocortisone 100 mg) were given before the induction of general anesthesia. Intraoperative neurophysiological monitoring, including motor evoked potential and somatosensory evoked potential and electromyogram (for cranial nerves III, IV, and VI functions) were used. The patient was then put in a supine position with the head of the operating table elevated by 20 degrees to facilitate venous drainage. The patient's head was turned to the right about 10 degrees, with the nasal bridge parallel to the floor. The two teams commenced surgery simultaneously.

The middle and inferior turbinates were outfractured. A large septal defect was confirmed and the sphenoid rostrum was deficient from previous surgery. The distorted anatomy made the approach difficult, but by sequentially identifying the bilateral carotid protuberances, the undersurface of tuberculum sellae, and both medial and lateral opticocarotid recesses, the sella was finally visualized and its extent verified with image guidance. The bony sella floor had been replaced by fibrous tissue and sphenoid mucosa. The tumor capsule was then opened in a cruciate fashion, followed by tumor debulking. The operating table was then turned 30 degrees to the right to facilitate the cranial surgeon's work (see **Operative Setup**).

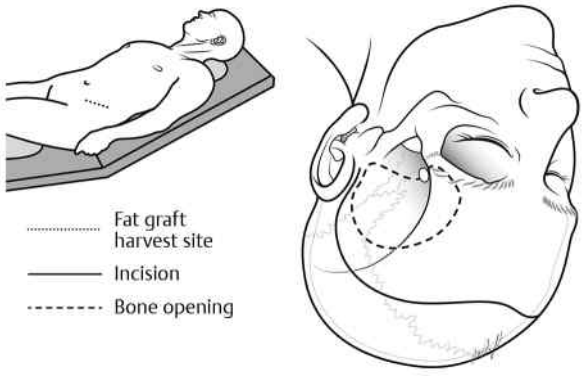
Operative Setup

Position: supine, head turned 10 degrees to the right.

Incision: reopening of the previous bicoronal incision and endonasal TS route

Bone opening: left pterional craniotomy

Durotomy: from distal dural ring toward the sylvian fissure



The cranial surgeon reopened the previous bicoronal incision, and the scalp flap was turned anteriorly after interfacial dissection. A left pterional craniotomy was made, and temporal squama was flattened to the floor of the middle cranial fossa. The frontal and temporal lobes were retracted epidurally with stay sutures. The sphenoid ridge was drilled flat. The meningo-orbital band connecting the outer border of superior orbital fissure and the frontotemporal dura was cut to gain access to the anterior clinoid (**Fig. 11.5**). Following a lateral orbitotomy, an extradural anterior clinoidectomy was performed with diamond drill. The drilled surface of the optic strut was waxed carefully to prevent any leak into the sphenoid sinus.

The clinoidal triangle was exposed by this stage. The outer layer of the lateral cavernous sinus wall was then dissected from the inner layer, starting anteromedial, and moving posterolaterally (**Fig. 11.6**). Further posteriorly, the maxillary (V2) and mandibular (V3) branches of the trigeminal nerve (cranial nerve V) as well as the gasserian ganglion were dissected off

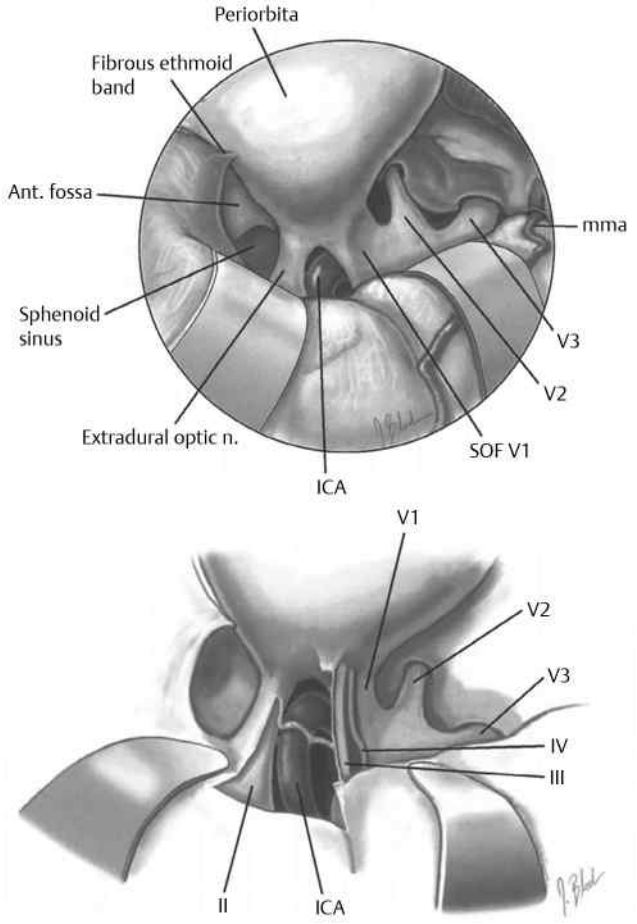


Fig. 11.5 The Dolenc approach to the cavernous sinus (right). After a pterional craniotomy, the sphenoid ridge was drilled flat. Lateral orbitotomy, anterior clinoidectomy, unroofing of the optic canal, and removal of the lateral wall of the superior orbital fissure (SOF) allowed the mobilization of the outer leaflet of the lateral wall of the cavernous sinus. (Reproduced from Harsh G, ed. Chordomas and Chondrosarcomas of the Skull Base and Spine. 1st ed. Thieme; 2003.)

the temporal base dura. The lateral cavernous sinus wall bulged out because of the tumor, and incision into the lateral wall was made between the trochlear nerve and V1 (Parkinson's triangle) (**Fig. 11.7**). The tumor within the lateral compartment was soft and readily removed by suction. To gain further exposure, the trochlear nerve had to be sacrificed as the dense fibrosis of the wall made its dissection impossible. The oculomotor nerve was gently retracted to medial side, while ICA and V1 were retracted laterally to clear the medial and posterosuperior compartments. Venous bleeding was encountered stemming from the cavernous sinus and it was stopped by packing. A small communication between the sella and the medial cavernous sinus wall was sealed with small pieces of subcutaneous fat graft harvested from the abdomen.

The intradural component of the cranial procedure began with dura opening along the sylvian fissure toward the optic canal. The distal dural ring was divided to expose the roof of the cavernous sinus. Intradurally, the tumor surrounded the oculomotor nerve over the roof of cavernous sinus and abutted the optic nerve and chiasm. It was dissected away from all these structures and removed. Thereafter, the capsule of the suprasellar tumor bulk was found medially which was pushed

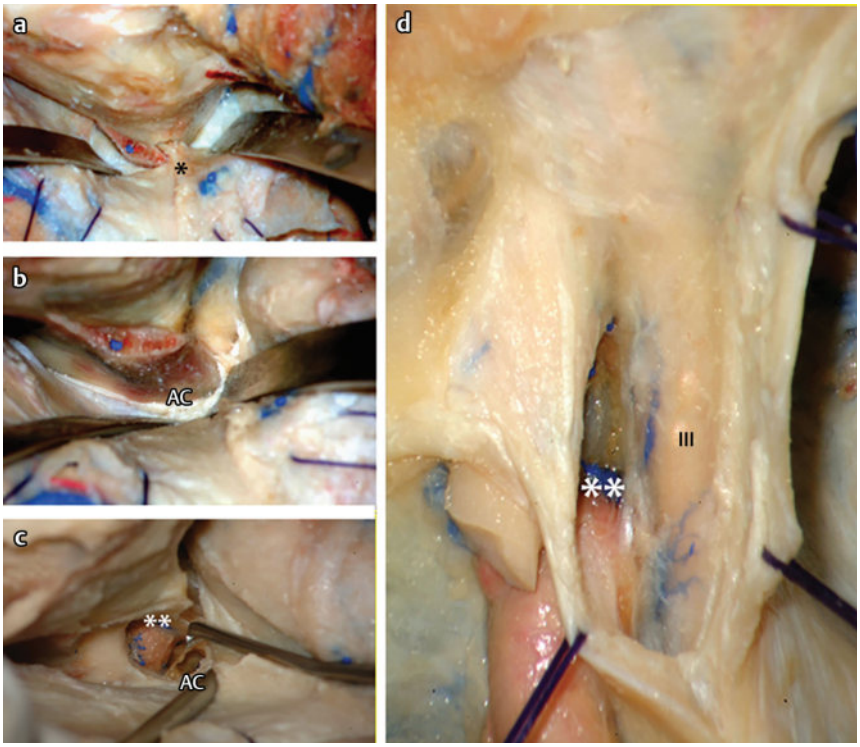


Fig. 11.6 Cadaveric dissection (right) demonstrating an extradural anterior clinoidectomy and mobilization of the lateral wall of the cavernous sinus (CS). (a) After lateral orbitotomy the meningo-orbital band (*) is encountered and cut; **(b)** this exposes the anterior clinoid process (AC) at its base; **(c)** after drilling the optic strut, the AC is fractured posterolaterally, exposing the clinoid segment of the ICA (**); **(d)** the lateral leaflet of the lateral wall of the CS (held by two lateral sutures) is mobilized away from the inner leaflet starting at the oculomotor nerve (III).

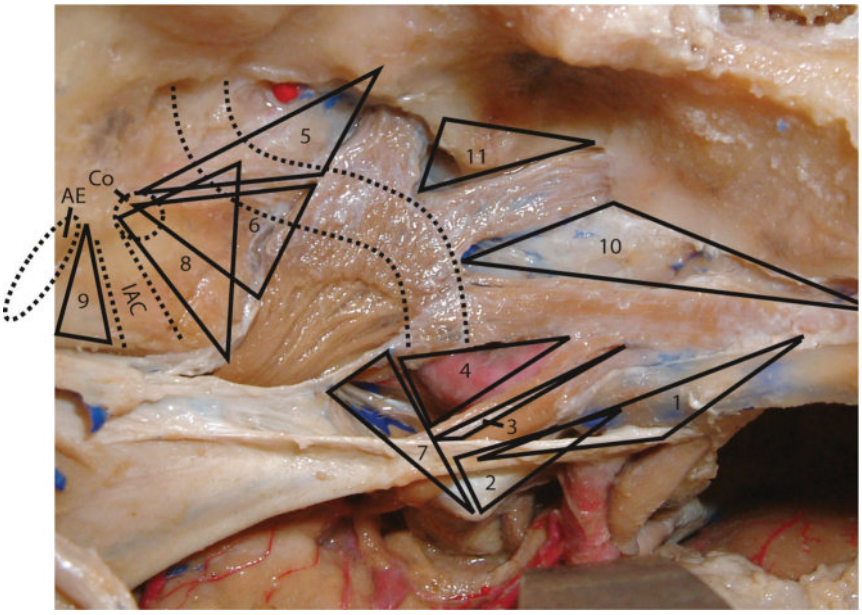


Fig. 11.7 Cavernous sinus triangles (left). 1, anteromedial triangle (Dolenc); 2, medial triangle (Dolenc, Hakuba); 3, superior triangle (Fukushima); 4, lateral triangle (Parkinson); 5, posterolateral triangle (Glasscock, Paullus); 6, posteromedial triangle (Kanzaki, Kawase); 7, posteroinferior triangle (Fukushima); 8, premeatal triangle (Day, Fukushima); 9, postmeatal triangle (Day, Fukushima); 10, anterolateral triangle (Mullan); 11, far-lateral triangle (lateral loop) (Dolenc). AE, arcuate eminence; Co, cochlea; IAC, internal auditory canal. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

gently toward the sphenoidal sinus for removal by the TS surgeon (**Fig. 11.8**). This resulted in an effective decompression of the optic chiasm, right optic nerve, bilateral supraclinoid ICAs and A2 branches. The pituitary stalk was preserved. Posteriorly, the Liliequist membrane was opened and the basilar artery (BA) was visualized. Luckily, the tumor dissected free from the BA quite readily and resection was completed by the TS surgeon as the tumor capsule descended down toward the sella.

Surgical repair of the skull base defect was conducted from below. Since the sella opening was small, we repaired it in multiple layers without vascularized flap. The sella itself was packed with fat graft under monitoring by the cranial surgeon to make sure it was sufficient but not overdone. Both nostrils were packed with Merocel. On the cranial side, the cranial dura was closed with temporalis fascia, and reinforced with fat graft and tissue glue. The craniotomy bone flap was replaced and secured in the usual manner. The previous VP shunt was removed and replaced by an external ventricular drain (EVD).

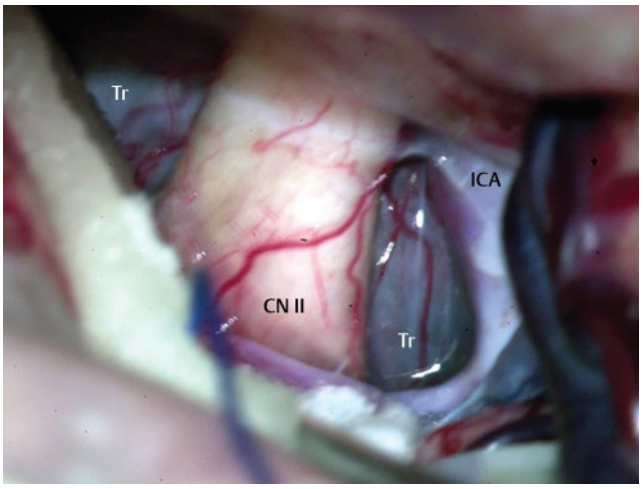


Fig. 11.8 Intraoperative image from cranial side under microscope. Residual tumor and capsule were seen under left optic nerve through the opticocarotid triangle. CN II, cranial nerve II; ICA, internal carotid artery; Tr, tumor capsule.

Surgical Pearls

1. It is important for the cranial and TS surgeons to work in a coordinated manner, especially when dealing with the suprasellar tumor portion. The cranial surgeon should avoid breaching the tumor capsule, while always pushing tumor into the sella for removal. Meanwhile, the TS surgeon should stay within the sella and not venture beyond. This cycle of "dissect, push down, resect" should be conducted repeatedly with good communication between the two teams. The key concept in two-team simultaneous surgery is that each team assists the other. This is not possible if the two operations were staged.
2. Occasionally, the opening at the sellar diaphragm may be too small to allow the delivery of tumor into the sella. The TS surgeon may need to enlarge the opening by making relieving incisions on the diaphragm.
3. The two teams need to compromise on how much the patient's head needs to be turned relative to the horizon. It should be turned more toward the TS surgeon to facilitate orientation at the initial stage. Once the sella has been safely entered, the operating table can be turned to facilitate the cranial surgeon's work.
4. Stereotactic image guidance is essential for redo TSS where normal anatomy may be distorted.

■ Aftercare

Our patient had a fair postoperative recovery. Her left eye remained blind and the right eye was able see hand movement only. She also had a new onset of partial left cranial nerve III palsy and transient paresthesia over the left V1 dermatome. Continuous cerebrospinal fluid (CSF) drainage through the EVD was maintained to promote healing. Nasal pack of the EVD were both kept for 5 days. She had no CSF leakage and no evidence of hydrocephalus after the removal of EVD. Despite a transient episode of diabetes insipidus, she was discharged with only her preoperative cortisol and thyroid hormone replacement.

Postoperative MRI showed a near-total resection with residual tumor found anteromedial to the left cavernous ICA (**Fig. 11.9a**), but she declined another course of radiation. Presently at 3 years after surgery, there is no evidence of regrowth (**Fig. 11.9b**).

■ Potential Complications and Associated Management

CSF leakage is a known complication of TSS. Specific to this patient, there is the potential risks of leakage through the optic strut defect after clinoidectomy, and through the defec-

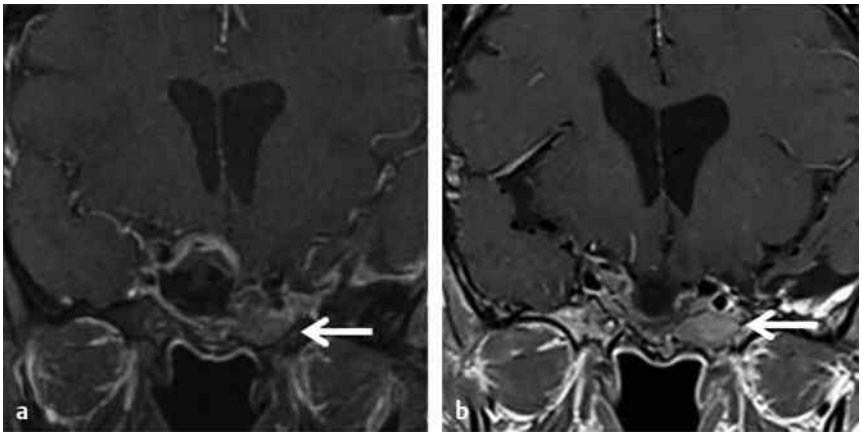


Fig. 11.9 Postoperative coronal MRI pituitary T1 sequence with gadolinium. (a) Three months after surgery. **(b)** Three years after surgery. Residual tumor (*arrow*) remained stable.

tive medial wall of the cavernous sinus into the sphenoid sinus. These areas should be carefully sealed off with bone wax, muscle, fat graft, and artificial sealants during surgery. CSF leakage should be managed aggressively with prophylactic antibiotics and CSF drainage, and definitive surgical exploration and repair if needed. Computed tomography (CT) cisternogram can prove invaluable in defining the site of leakage. Vascularized flaps may be needed in case of a refractory CSF fistula and permanent CSF shunting should be liberally considered.

Hormonal insufficiency is another common complication with diabetes insipidus being the most frequent after large-scale operations such as this. Monitoring urine output and serum sodium levels is critical for detection. Ophthalmoplegia and visual deterioration can occur anytime during the postoperative period, and the causes may include surgical injury, overpacking of sella, residual tumor, edema, and hemorrhage. One rarely discussed problem is vasospasm, which may occur if the tumor capsule was opened with significant hemorrhage within the subarachnoid space. The patients can present with a myriad of confusing symptoms. The prudent use of angiography in such a situation can lead to a quick diagnosis and proper aggressive treatment. Ideally, the subarachnoid space should be thoroughly irrigated before closing.

Perspective

Gabriel Zada

The authors of the previous section presented a very challenging case of a refractory and multiply recurrent nonfunctioning pituitary macroadenoma. The patient was first treated over two decades ago via a TS operation, and this was followed by radiation therapy early on. She was also treated with a subsequent interhemispheric craniotomy and several additional TS procedures.

Nonfunctioning pituitary adenomas are of course benign lesions. However, when they have recurred several times despite fractionated radiation therapy, they can be challenging tumors that behave more like locally malignant tumors. For the selection of a treatment plan, which might include surgery and nonsurgical therapies, the salient facts of this case included the following: (1) the patient was nearly blind bilaterally; (2) she had hypopituitarism; (3) the tumor was irradiated years ago; (4) she has had numerous TS operations in the past, making this approach less favorable; (5) she had a VP shunt.

For such a complex situation, the histopathological identity and biological behavior of the tumor cells are important to consider. For example, if the immunostaining data from surgery show a silent adrenocorticotropic hormone adenoma, the therapeutic strategy moving forward may be different from a null-cell adenoma. Similarly, the MIB-1 labeling index could also impact the decision on further treatment.

I agree with the previous authors that the surgical goal for this complicated patient should be maximal safe tumor resection, with the additional aim of tissue acquisition for molecular profiling and immunohistochemistry analysis. Given the Knosp invasion score, a total resection would be unrealistic without new and significant neurological consequences. The expected residual tumor after surgery would presumably be a favorable target for subsequent radiosurgery, which should be safe because the patient was already nearly blind in one eye, had hypopituitarism, and irradiated many years ago.

Although this general scheme would involve surgery using multiple modalities, what made the actual plan unique was the decision to perform transcranial and endonasal surgery simultaneously. I present another case for comparative discussion.

■ Case Presentation

A 43-year-old man presented with vision loss and diminished libido. He was found to have a profound bitemporal hemianopsia and low serum testosterone level. His MRI showed a large nonfunctioning pituitary macroadenoma with suprasellar extension and compression of the optic apparatus (**Fig. 11.10a, b**).

■ Anatomical and Therapeutic Considerations

The choice of surgical approach for this hypogonadotropic man may include an open craniotomy, endoscopic endonasal approach (EEA), or a combined approach. For first-time operations, I prefer to start with a resection from below (i.e., EEA) in an attempt to avoid a craniotomy, and deliver the whole tumor from below if possible. Although a concurrent craniotomy was possible in this same setting to facilitate the removal of the suprasellar component, I typically prefer to start with the EEA, and then if necessary, use the craniotomy at a later stage. This minimizes OR and anesthesia time for the patient, allows the patient time to recover between stages, and minimizes the risk of postoperative complications, such as CSF leaks and meningitis. Separating the stages also permits imaging studies during the interval, which could potentially reveal that the residual tumor after the EEA is smaller enough to treat with radiosurgery instead of a craniotomy.

The next step in planning is to decide on which type of EEA to use. Compared to a typical endoscopic endonasal TS approach, an extended transtuberculum approach offers a higher likelihood of achieving decompression of the optic apparatus, and it was therefore the best choice for this patient with a big tumor whose vision was profoundly impaired.

■ Description of the Technique and Clinical Outcome

The patient underwent an extended endonasal transtuberculum approach, and during surgery, the tumor was found to be very vascular and firm. For this reason, residual suprasellar tumor was knowingly left behind for safety concerns. Reconstruction was achieved via an autologous fascia lata graft and rotation of a pedicled nasoseptal flap. A postoperative MRI (**Fig. 11.10c, d**) showed residual tumor in the suprasellar cistern that had descended slightly, but was still very close to the optic chiasm and nerves. Although fractionated stereotactic radiosurgery was an option for him, the close proximity of the tumor to the visual apparatus made this suboptimal. Furthermore, given his young age and the questionable durability of the effects of radiosurgery, the patient and surgical team agreed that additional surgical resection was the better plan.

Although a redo endonasal approach or a combined endonasal and craniotomy approach were possible options, a transcranial approach using the anterolateral corridor was chosen because the endonasal route had already been closed with a pedicled nasal–septal flap. Furthermore, the tumor was known to be firm and vascular, and these aspects may be better controlled from a transcranial route. A pterional craniotomy is the standard opening for the anterolateral corridor, but since a supraorbital eyebrow approach provides adequate exposure, the decision was made to use this less invasive technique (**Fig. 11.11**).

The result of the right supraorbital eyebrow keyhole approach was a gross total tumor resection (**Fig. 11.10e, f**). The patient's vision improved after the second surgery. His hypogonadism remained, but he did not develop any further endocrine issues.

■ Commentary

This particular case highlights a successful strategy employing a staged operation for a complex pituitary macroadenoma. Both

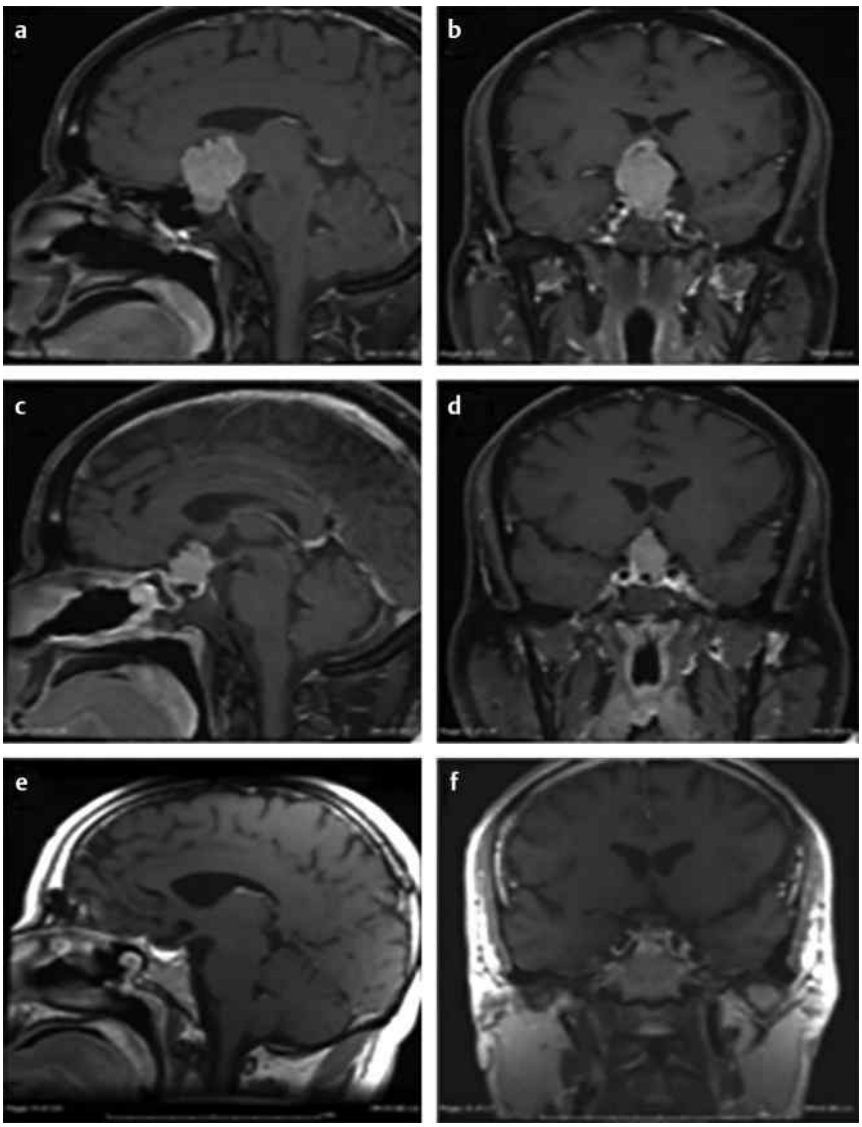


Fig. 11.10 MRI images of the 43-year-old patient described in this chapter. (a, b) Preoperative images; **(c, d)** after transsphenoidal surgery; **(e, f)** after supraorbital keyhole approach.

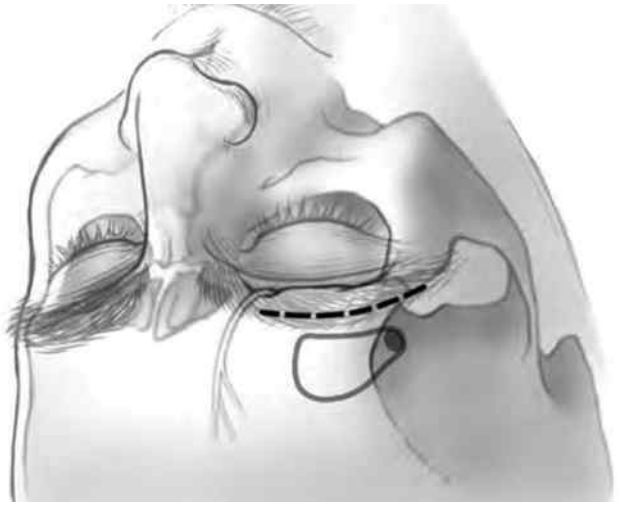


Fig. 11.11 Right supraorbital eyebrow keyhole approach. Schematic illustration showing the relative locations of the incision and bone flap in this approach. (Reproduced from Teo C, Sughrue M, eds. Principles and Practice of Keyhole Brain Surgery. 1st ed. Thieme; 2015.)

cases in this chapter needed a multidirectional approach, combining EEA with transcranial surgery. The difference between the examples was whether the surgical modalities were used simultaneously or in a staged fashion.

As the authors of the previous section pointed out, there are some benefits for performing the simultaneous approach. A concurrent above-and-below approach theoretically offers improved views of the lesion from multiple angles, and allows a dissect-from-above and deliver-from-below strategy. The ability to dissect the tumor from surrounding neurovascular structures, such as the optic nerves, vessels of the circle of Willis, and perforator vessels, may be accomplished more safely from above, thereby facilitating more aggressive tumor removal from below.

However, the simultaneous above-and-below technique also has significant disadvantages, and in particular, increases the risk of postoperative infection. In fact, complications in *either* phase of the operation, whether "above" or "below," may potentially jeopardize *both* approaches, and threaten the repair of the skull base at the end of the operation, which separates the two compartments.

These weaknesses do not apply to the staged strategy, as the patient is given time to heal between the stages. Staging also

has several additional advantages which ultimately makes it my preference. The residual tumor after the first stage may in fact be small enough to treat with radiosurgery, making a transcranial surgery unnecessary. In addition, even with two separate operations, the total anesthesia time may still be less than that required for the simultaneous above-and-below operation.

Conclusion

The authors of the previous section presented a complex case, and they achieved a radical resection using the combined, simultaneous approach, while avoiding postoperative rhinorrhea or infection. However, the patient did develop a third cranial palsy and has persistent near-total vision loss. A staged surgical strategy is worthy of consideration as an alternative. However, ultimately, successful long-term tumor control of this patient and similar ones will rely on additional adjunctive multimodality approaches, including radiosurgery and medical/targeted therapy.

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12 Posterior Clinoid

Hiroki Morisako, Takeo Goto, and Kenji Ohata

Keywords: petrosectomy, partial labyrinthectomy, sphenobasal vein, eyebrow keyhole, endoscope, retroclival

■ Case Presentation

A 39-year-old previously healthy man presented to a community hospital with a 2-month history of dysarthria and gait disturbance. The symptoms were gradual in onset, and the family had noticed he had some difficulty swallowing during the last month. On neurological examination, he had truncal ataxia and was partially hemiparetic on the left side. His magnetic resonance imaging (MRI) showed a large mass region at posterior clinoid process (**Fig. 12.1**).

Questions

1. Given the unique location of the mass, what is the differential diagnosis of the MRI finding?
2. What kind of examination is required before the next step in surgical management?

■ Diagnosis and Assessment

The patient's blood work was normal for routine investigations as well as anterior pituitary hormones. Neither his personal nor family medical history was remarkable. His MRI revealed a well-defined, 5 cm in diameter, solid mass that was isointense on T1-weighted image, hyperintense on T2-weighted image, and homogeneously enhanced in the intra- and suprasellar portions without calcification. The lateral and third ventricles were dilated. The differential diagnosis for the tumor includes meningioma and a fibrotic type of mesenchymal tumor.

Exceedingly rare (< 1% all central nervous system tumors), these mesenchymal tumors include a histological spectrum previously classified separately as meningeal solitary fibrous tumor and hemangiopericytoma. Peak incidence occurs in the fourth to fifth decade of life, and males are affected slightly more frequently than females. Most of these tumors are dural-based, and skull base, parasagittal, and falcine locations are especially common. On MRI, the tumors are isointense on T1-weighted images, with high or mixed intensity on T2-weighted images. On gadolinium administration, enhancement is variable. Dural contrast enhancement at the periphery of the lesion and flow voids may be observed. In other words, these tumors are nearly indistinguishable from meningiomas on MRI features, and the main difference is that meningiomas are more common.

In preparation of surgical intervention, digital subtraction angiography (DSA), computed tomography angiography (CTA) and venography (CTV) were obtained on our patient to assess the position of the feeding arteries of the tumor, the venous system and its relationship with the tumor mass. CTA showed superior displacement of the A1 segment of the right ante-

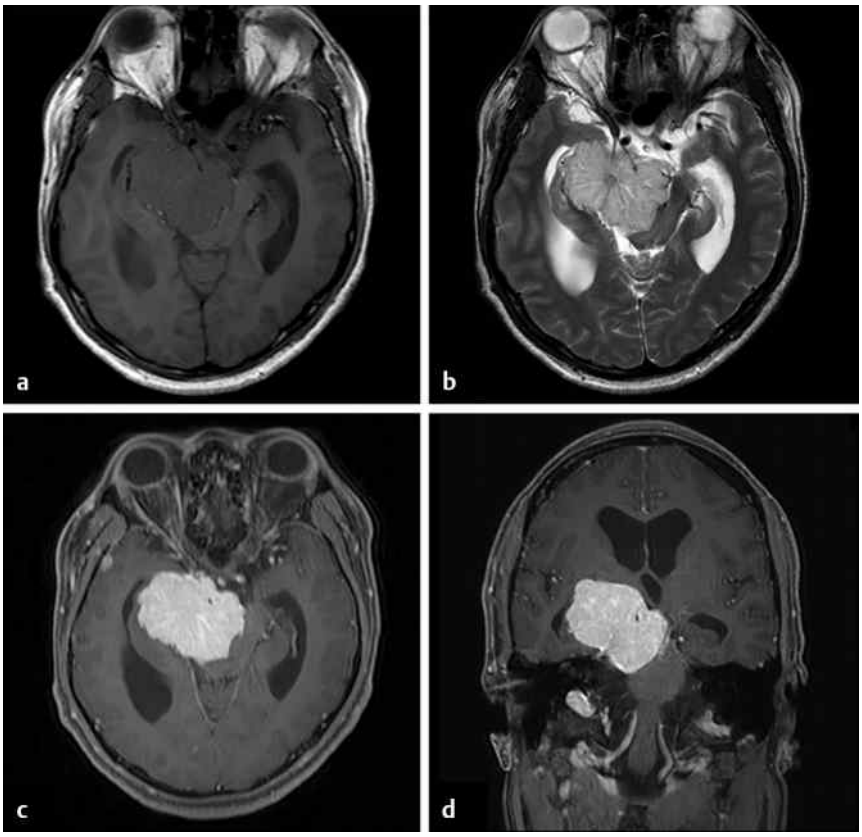


Fig. 12.1 **Preoperative MR images.** **(a)** T1-weighted axial image; **(b)** T2-weighted axial image; **(c)** T1-weighted axial image with gadolinium administration; **(d)** T1-weighted coronal image with gadolinium administration. These showed a large mass at the right posterior clinoid process.

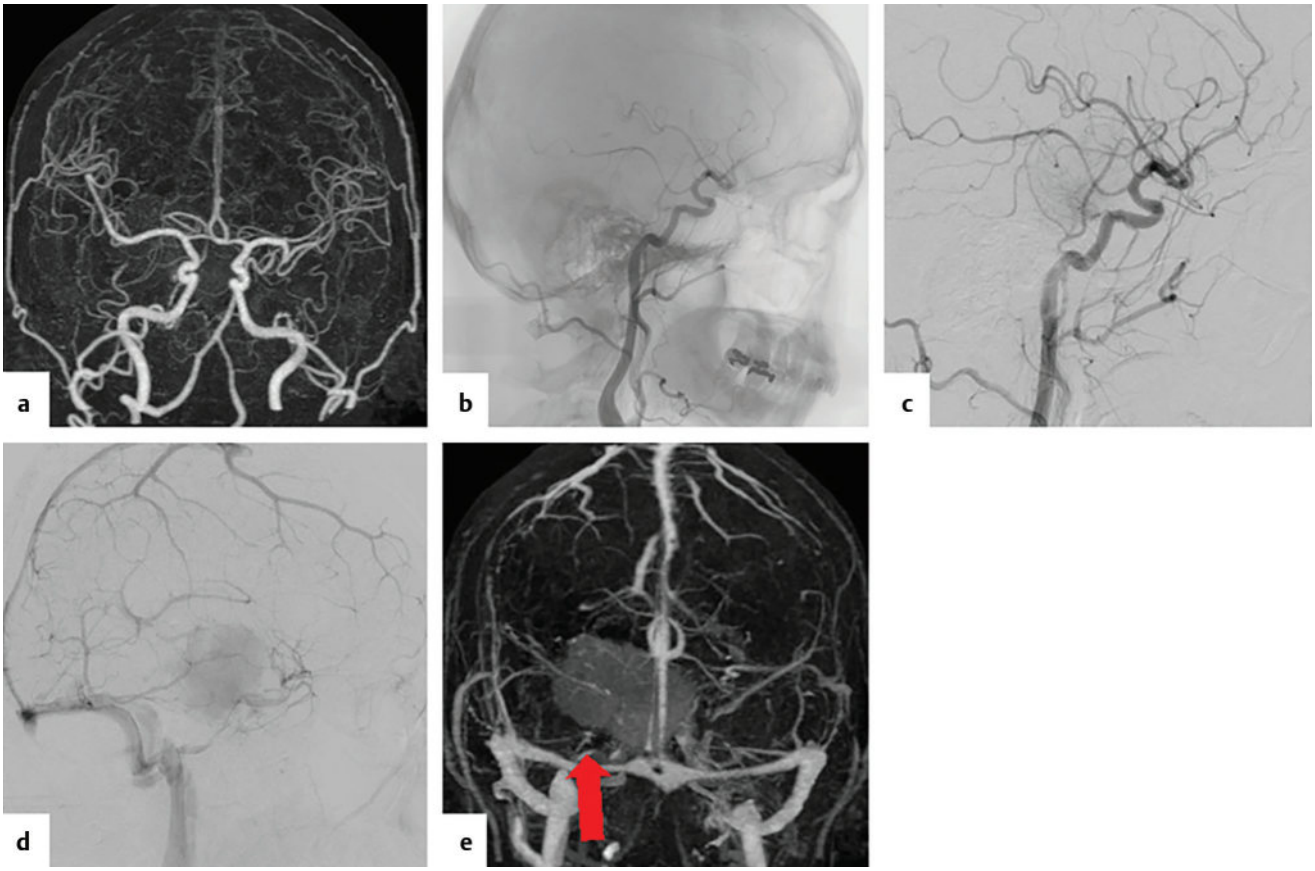


Fig. 12.2 Preoperative vascular imaging of the case. (a) CT angiogram showed displacement of the anterior circulation on the right side. **(b, c)** Arterial phase of digital subtraction angiography showed the tumor "blush" and the displacement of the anterior cerebral artery, middle cerebral artery, and communicating segment of the right internal carotid artery. **(d)** Venous phase of the right common carotid artery injection demonstrated that the venous plexus around foramen ovale was not dilated. **(e)** CT venography showed a large superior petrosal vein connecting with the superior petrosal sinus (*red arrow*).

rior cerebral artery (ACA) and the M1 segment of the right middle cerebral artery (MCA) (**Fig. 12.2a**). DSA of the right common carotid artery demonstrated the tumor "blush" and the displacement of the communicating segment of the right internal carotid artery (ICA), ACA, and MCA (**Fig. 12.2b**). The tumor was fed by the right meningohypophyseal trunk from the ICA and the right middle meningeal artery from a maxillary artery (**Fig. 12.2c**). Venous phase of DSA showed the venous drainage of right temporal lobe via the vein of Labbé and superficial middle cerebral vein (sylvian vein). Unlike some other tumors of this region, the venous plexus around the foramen ovale was not dilated (**Fig. 12.2d**); however, the CTV demonstrated a large superior petrosal vein draining to the superior petrosal sinus (**Fig. 12.2e**).

■ Anatomical and Therapeutic Considerations

The radiographic findings were consistent with a meningioma of the posterior clinoid process (PCP). The PCP is located posterolaterally to the pituitary stalk, posteromedially to the oculomotor nerve at its entrance into the cavernous sinus, and just behind the communicating segment of the ICA. Meningioma arising from the PCP often displaces the pituitary stalk anteriorly, the oculomotor nerve laterally or inferolaterally, and the communicating segment of the ICA and its branches superiorly or anteriorly. If the tumor expands significantly anteriorly, optic nerve and chiasma will be shifted superiorly or anterosuperiorly, leading to visual disturbance. A large tumor can lead to hydrocephalus as it compresses the third ventricle.

In the radiological examinations, attachment of the tumor at the PCP is very small and dural tail sign is not usually detected. Some posterior clinoidal meningiomas can be confused with other parasellar tumors or even anterior clinoidal meningiomas. The direction in which the ICA is displaced can offer clues to differentiate between anterior and posterior clinoidal tumors. The artery is shifted posteriorly when the tumor arises from the anterior clinoid, whereas the shift is anterior for PCP meningiomas.

Options for Approach

Since this patient is young and otherwise healthy, and most meningiomas are slow-growing tumors, the decision for surgical removal was made with an aim on safe maximal resection. Posterior clinoidal meningiomas are very rare, and there are few reports and even less agreement on their surgical management. However, in general terms, the removal of PCP meningiomas requires the following five steps: (1) devascularization of

the tumor at posterior clinoid process, (2) location of the oculomotor nerve, (3) dissection of the tumor from the ICA and its branches such as the anterior choroidal artery, (4) dissection from the pituitary stalk and the hypothalamus, (5) dissection from the optic chiasma and nerve. Some surgeons advocate staged, multidirectional approaches to accomplish these steps, for example, with the first two performed through a standard lateral suboccipital approach and the remaining steps through a pterional approach. Combination of these two approaches may be suitable for relatively small and soft posterior clinoidal meningioma.

Another option for approach to the lesion is the transcavernous–transsellar approach, which can accomplish all five steps in one stage. However, the disadvantage of this approach for a PCP meningioma is that, with the ICA and its branches shifted anteriorly, they can obstruct the surgeon's line-of-sight in the anterolateral corridor. This increases the surgical risk, as the tumor is removed through a narrow space between perforators of ICA. Moreover, peeling off the outer layer of the lateral wall of the cavernous sinus sometimes obstructs venous return from sphenoparietal sinus. Early detection of the oculomotor nerve is also difficult in this approach.

If the anterolateral corridor is suboptimal because of an obstructed view by the ICA and a long reach to the PCP, then logically, shifting the approach laterally may be the solution. A middle fossa craniotomy coupled with an anterior petrosectomy would provide a short, direct approach to the main bulk of the tumor, but the surgeon's line-of-sight for this approach is directed downward, toward the posterior fossa. In contrast, the main bulk of the PCP meningioma is pointed upward and slightly anteriorly from the level of the middle fossa floor, and thus, an operative approach that has an inferior-to-superior, posterior-to-anterior working trajectory is best suited for this tumor.

Approach of Choice

The presigmoid transpetrosal approach utilizes the posterolateral corridor, and has the requisite upward and anteriorly-directed trajectory that is ideal for PCP meningiomas. In fact, during the last 20 years, we have encountered six cases of posterior clinoidal meningiomas and successfully treated them using the lateral corridor with a presigmoid transpetrosal approach. This approach can be tailored for PCP meningiomas of any size and firmness. The main caveat is, in a case where venous return mainly passes through the sphenobasal vein and venous plexus around the foramen ovale, dural elevation from the middle fossa floor must be limited to preserve the venous return. In our patient, we planned a presigmoid transpetrosal approach with a partial labyrinthectomy to preserve his hearing.

Questions

1. How would you position the patient for a presigmoid transpetrosal approach?
2. Which semicircular canal(s) would you preserve during the partial labyrinthectomy?

Description of the Technique

Position and Skin Incision

The patient was placed in a semiprone park bench position. The head was fixed using a three-point fixation with the head rotated and vertex down to keep the temporal side of the head in the horizontal plane. The skin incision started at the upper margin of the zygomatic arch anterior to the tragus, turned 2 to 3 cm above the ear, and then descended behind the posterior margin of the mastoid process. After raising the skin flap, a temporofascial pericranial flap was harvested based on the sternocleidomastoid muscle pedicle, which was used for the dural reconstruction at the end, to prevent postoperative cerebrospinal fluid (CSF) leakage (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral

Craniotomy: temporal + lateral suboccipital

Modifiers: posterior petrosectomy, partial labyrinthectomy

Craniotomy

Temporo-occipito-suboccipital craniotomy was performed prior to mastoidectomy. Seven burr holes were used, placed at specific landmarks to avoid injury to the sigmoid sinus. The locations were as follows: (1) the asterion, (2) the intersection of the supramastoid crest with the squamous suture, (3) the mastoid emissary foramen, (4) the root of zygoma, (5) the transverse sinus, (6, 7) the anterior and posterior aspect of the temporal bone, respectively. Following the standard temporo-occipito-suboccipital craniotomy, the outer table of the mastoid process was split to prevent the postoperative cosmetic deformity. The sigmoid sinus was carefully exposed down to its short horizontal segment (see **Operative Setup**).

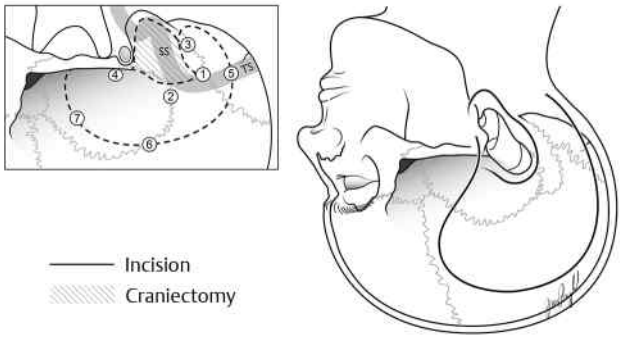
Operative Setup

Position: semiprone, park bench

Incision: "U-shaped" over temporal region

Bone Opening: "L-shaped," temporal + lateral suboccipital

Durotomy: subtemporal and presigmoid



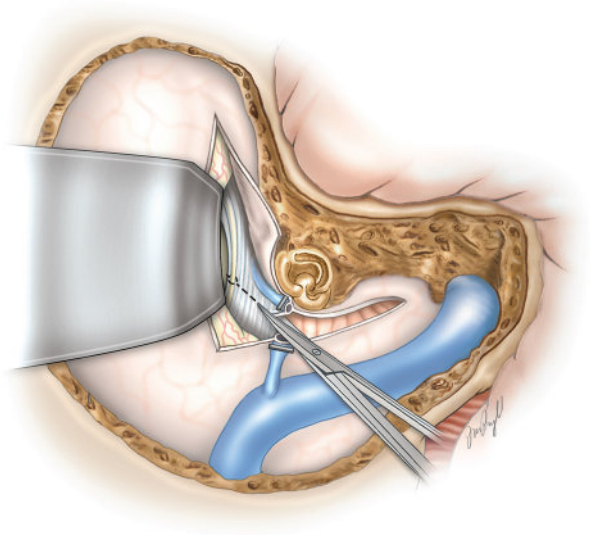


Fig. 12.3 Illustration showing the operative field after the posterior petrosectomy. The superior petrosal sinus was ligated and transected at a point *anterior* to its connection with the superior petrosal vein in order to preserve venous drainage.

Extent of Posterior Petrosectomy

A retractor was used to protect the sigmoid sinus, and the petrosectomy was performed. The posterior and superior semicircular canals were left untouched to preserve hearing function. The petrous ridge was removed up to the level of internal auditory meatus. The petrous apex was resected to expose the entire course of the superior petrosal sinus (**Fig. 12.3**). During the extradural portion of the procedure, one should avoid unnecessary elevation of the dura off of the middle fossa floor, which can disrupt the venous drainage.

Incision of Dura and Tentorium

The petrosal dura was opened, leaving 5-mm dural fringe on the anterior margin of the sigmoid sinus and on the inferior margin of the superior petrosal sinus. The subtemporal dural incision was extended as far anterior as possible. Its medial extent coursed along the posterior margin of the gasserian ganglion. The superior petrosal sinus was transected at a point *anterior* to its connection with the superior petrosal vein in order to preserve venous drainage (**Fig. 12.3**). The tentorium was divided from the point of the transaction of the superior petrosal sinus to the incisura, in a course directed posterior to the entry of the trochlear nerve into the tentorial edge (**Fig. 12.4a**).

Tumor Removal

Gentle elevation of the temporal lobe provided a wide exposure of the tumor above and below the trochlear nerve (**Fig. 12.4**). At the early stage of the operation, coagulation of the feeding arteries from the PCP was safely accomplished (**Fig. 12.5a**). After internal debulking of the tumor, the superior cerebellar artery (SCA) was found in the prepontine cistern (**Fig. 12.5b**). Tumor was

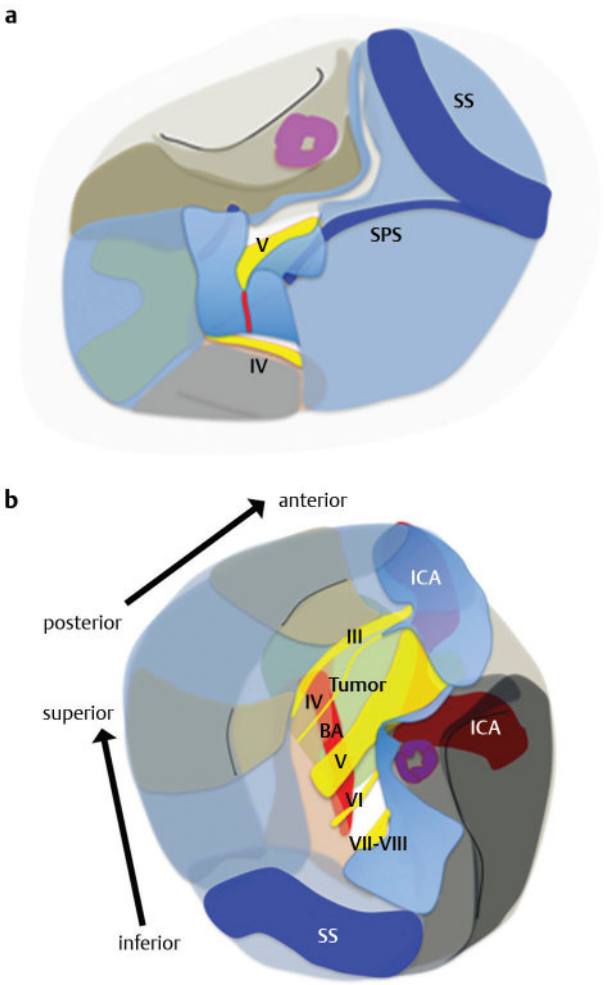


Fig. 12.4 Illustration demonstrating surgical steps in presigmoid transpetrosal approach (right side). (a) The tentorium was cut behind the dural entrance of the trochlear nerve (*red line*). SS, sigmoid sinus; SPS, superior petrosal sinus; IV, trochlear nerve; V, trigeminal nerve. **(b)** The wide exposure and unique posterior-to-anterior and inferior-to-superior projection for posterior clinoid area was obtained. BA, basilar artery; ICA, internal carotid artery; SS, sigmoid sinus; artery; III, oculomotor nerve; IV, trochlear nerve; V, trigeminal nerve; VI, abducens nerve; VII, facial nerve; VIII, acoustic nerve.

dissected away from the distal portion of the right SCA and brainstem (**Fig. 12.5c**). From a posterolateral vantage point, tumor was also safely removed from the ICA and its branches, which were displaced anterior-superiorly. At the end, a near-total resection of the tumor was achieved, as a small residual tumor was adherent to the right oculomotor nerve (**Fig. 12.5d**).

Closure

All opened mastoid air cells were sealed with abdominal fat soaked in fibrin glue. The mastoid and petrous portions of the temporal bone, as well as the dural defect, were entirely covered with the harvested temperofascial pericranial flap. The tempo-ro-occipito-suboccipital bone flap and the outer table of the mastoid process were replaced and fixated with titanium miniplates.

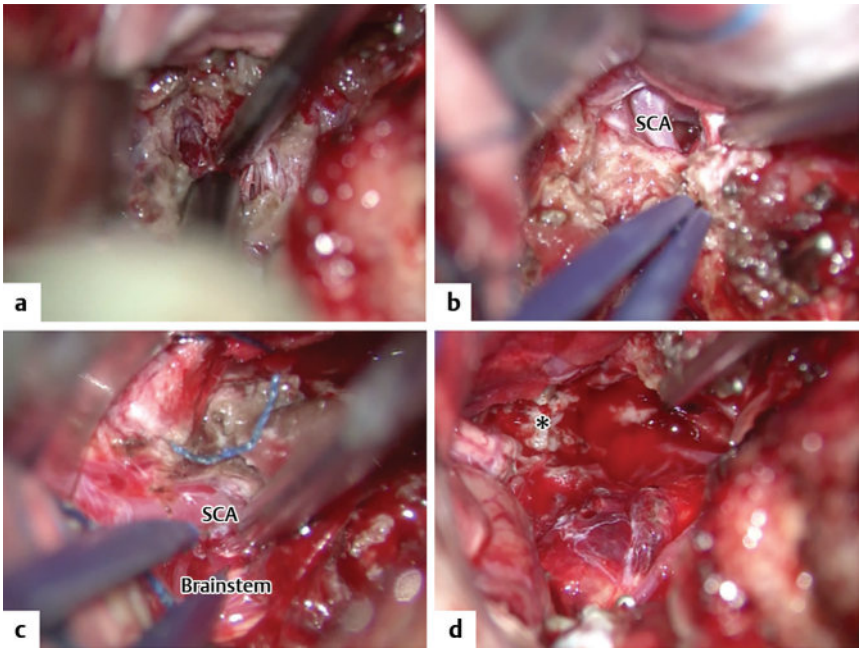


Fig. 12.5 Intraoperative images of the case. (a) After cutting of the superior petrosal sinus and opening of the Meckel's cave, internal debulking of the tumor was performed. **(b)** Proximal portion of the right superior cerebellar artery (SCA) was exposed. **(c)** Distal portion of the right SCA and brainstem was dissected from the tumor. **(d)** Near-total resection of the tumor was achieved with a small residual tumor along the right oculomotor nerve. SCA, superior cerebellar artery; *, residual tumor.

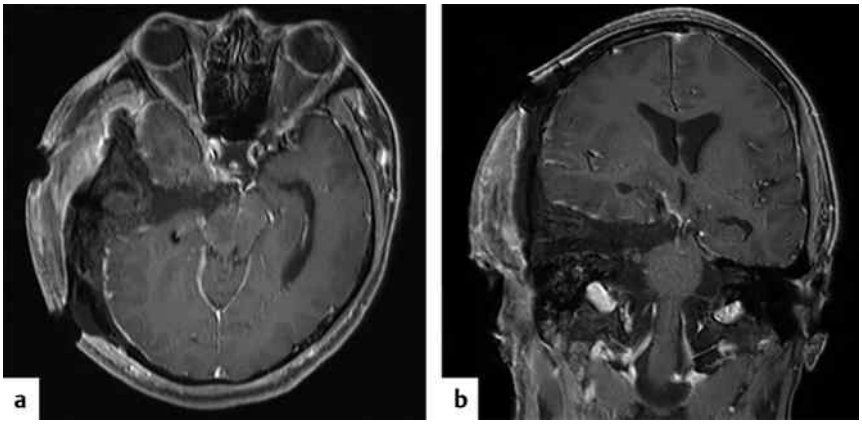


Fig. 12.6 Postoperative MR images. (a) T1-weighted axial image with gadolinium administration and **(b)** T1-weighted coronal image with gadolinium administration showing a near-total resection.

Surgical Pearls

Careful attention must be paid to the venous drainage to avoid complications:

1. To complete the approach to the tumor, the tentorium must be incised all the way to the incisura. In order to do this, the superior petrosal sinus must be ligated. For patients in whom preoperative images show an unusually dilated superior petrosal vein, it is critical to ligate the sinus *anterior* to the junction with the superior petrosal vein to preserve the flow from the vein to the sinus and then into the sigmoid/transverse sinus system.
2. Also important is the preoperative definition of the location of the tumor and its relationship to the sphenobasal vein, venous plexus around the foramen ovale, and superior petrosal vein. In the case where venous return passes mainly through the sphenobasal vein and venous plexus around the foramen ovale, dural elevation from middle fossa floor must be restricted to preserve those venous returns.

■ Aftercare

The patient did well after his surgery with minor complaints of transient right oculomotor palsy and left hemiparesis. These symptoms improved completely within 3 months. On MRI, 95% of the tumor was removed (**Fig. 12.6**). The histopathological diagnosis from the resection was meningothelial meningioma. MIB index (Ki-67) was less than 1%. The patient showed communicating hydrocephalus 1 month after tumor resection, and he underwent ventriculoperitoneal (VP) shunt placement. The patient was discharged 1 month after the VP shunt. There was no regrowth of the residual tumor during 18 months of follow-up, and he was able to continue in his previous occupation (**Fig. 12.7**)

■ Conclusion

Posterior clinoidal meningiomas are very rare, and to our knowledge, there is no literature detailing their surgical

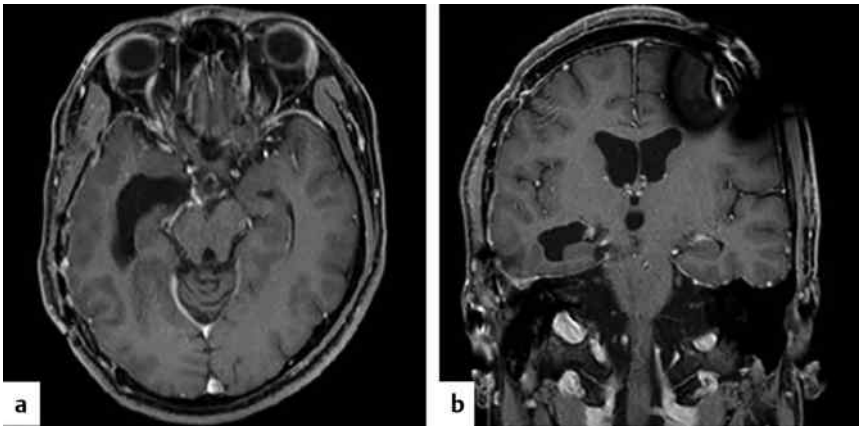


Fig. 12.7 MR images at the latest follow-up. **(a)** T1-weighted axial image with gadolinium administration and **(b)** T1-weighted coronal image with gadolinium administration showing no discernable trace of the tumor.

outcome except a case report. Nakamura et al reported a case of posterior clinoidal meningioma successfully resected in a two-stage operation combining lateral suboccipital approach and pterional approach. Dolenc emphasized rareness of the tumor. In his series of meningiomas of central skull base treated in the last 20 years, he had only six cases, which represent 0.7% of all lesions. He used the transcavernous–transsellar approach and removed the tumors completely without additional deficits except transient oculomotor nerve palsy.

In the last 20 years, we have encountered seven cases of posterior clinoidal meningioma. The patients included three males and four females, who ranged in age from 39 to 67 years (mean 52.6 years). The mean tumor diameter was 45.3 mm (range 32–55 mm). Two tumors were totally removed, and three had near-total resections by a presigmoid transpetrosal approach. One large tumor which had significant anterior extension was near-totally resected with a combination of transpetrosal approach and orbitozygomatic approach. One fibrous tumor was partially removed to avoid surgical complications. Five patients experienced transient oculomotor nerve palsy just after operation. Visual disturbance and cerebral infarction from the injury of perforators were avoided in all cases.

In our experiences, the presigmoid transpetrosal approach can be utilized for any variations of the posterior clinoidal meningioma. This approach also provides excellent visualization of the inferior surface of the chiasma, pituitary stalk, and hypothalamus. The limitation of the technique is the optic nerve; if a tumor extends anteriorly under the optic nerve, this portion will not be reachable. In this situation, an orbitozygomatic approach should be added to address this part of the tumor.

Perspective

Charles Teo and Steven Carr

■ Introduction

Tumors arising from the posterior clinoid are among the most difficult tumors to resect in neurosurgery, and several factors account for this challenge. First, the posterior clinoid is arguably the deepest part of the skull base, and standard approaches come with the challenge of a "long reach," with several important structures potentially obscuring the view. Second, tumors in this region can be in contact with or marshal blood supply from multiple intracranial vascular supplies, including both anterior and posterior circulations. Third, tumors can associate themselves with multiple cranial nerves entering the cavernous sinus (most notably the oculomotor nerve), and adjacent to it (i.e., the optic nerves). Fourth, anatomical variation in both the tumor growth trajectory and the patient's own normal anatomical structures can produce barriers to exposure. And lastly, tumors which have significant upward growth into the brain can be intimately associated with deep cerebral structures and difficult to dissect downward into view.

As the authors pointed out in the preceding section, debate exists about whether to treat large posterior clinoidal meningiomas as one mass, which can be visualized and removed from one massive approach, or use two approaches, each providing safe and reliably reproducible access to different tumor components. We subscribe to the latter philosophy, advocated in a similar form by Samii and Nakamura, and propose that the principles of keyhole surgery make this tumor manageable through a combination of an endoscopic-assisted keyhole supraorbital eyebrow approach, and a keyhole retrosigmoid craniotomy, in a single operative setting.

■ Case Presentation

A 40-year-old man presented with a 6-month history of experiencing unusual burning smells and a 2-month history of left-sided headaches. The rest of his medical history was unremarkable. An MRI scan demonstrated a probable meningioma arising from the left posterior clinoid and extending inferiorly to the upper third of the clivus (**Fig. 12.8**). His neurological examination was normal.

■ Anatomical and Therapeutic Considerations

Although in selected patients, especially the elderly with multiple comorbidities, a conservative subtotal resection may be preferable, in this young and healthy man the goal of surgery was curative safe resection. After the establishment of the surgical goal, selection of the ideal approach depends on the vascularity of the tumor, long axis of the lesion, compartments involved, experience and skill set of the surgeon, potential approach-related morbidities, anatomy of the skull base and underlying nasal sinuses, handedness of the patient, and several other more minor considerations.

Options for Approach

Approaches may be divided into endonasal and transcranial. Transcranial approaches may be further divided into microsurgical or microsurgical–endoscopic-assisted. Transcranial should also be divided into single or combined approaches. As discussed in detail in the previous section, the transpetrosal approach can be a useful direct route to tumors in this region. However, for left-sided lesions such as ours, this approach involves significant risk to the dominant temporal lobe. Drilling the petrous bone is time consuming and a laborious endeavor not routinely practiced by most neurosurgeons. It also runs the risk of damage to the ICA, facial nerve, as well as to hearing

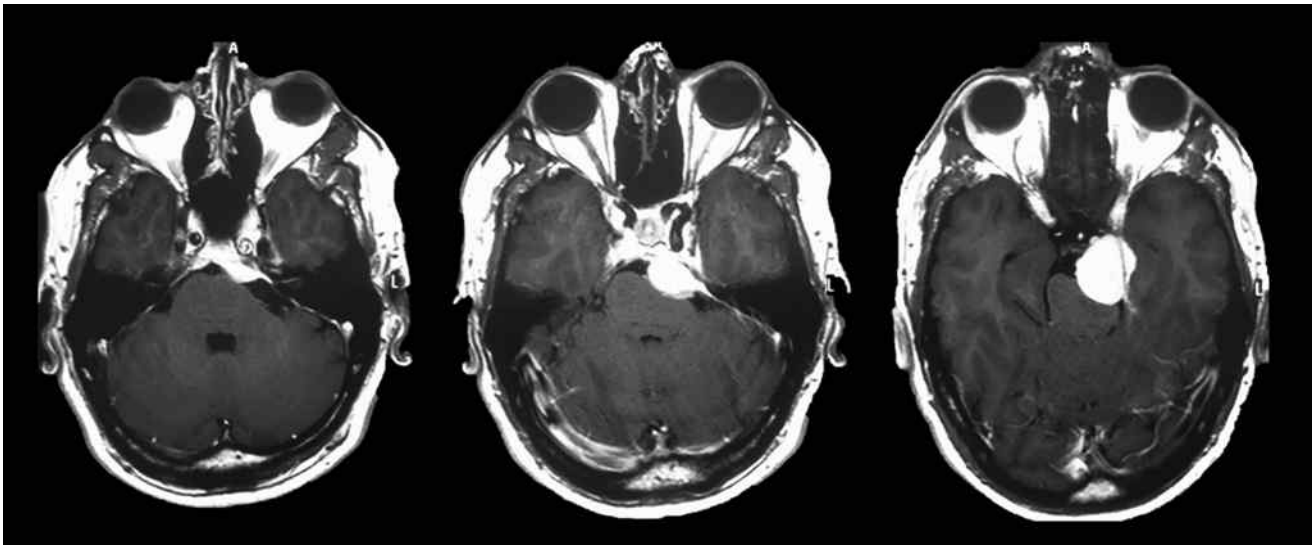


Fig. 12.8 Preoperative MRI. Axial views demonstrating a meningioma which originated from the left posterior clinoid process and extended inferiorly to the upper third of the clivus.

function. Finally, exposure to the MCA is limited through the transpetrosal approach, and dissection of a tumor which involves the MCA is performed with a greater margin for safety from a supratentorial approach.

A pterional craniotomy is one option that would enable safe dissection of supratentorial arteries, and paired with a retrosigmoid approach, would provide sufficient access to the posterior clinoidal meningioma. However, we do not favor this option because of the associated devascularization of the temporalis muscle, unnecessary frontal lobe exposure, and the bulky musculocutaneous flap which reduces the surgeon's visualization of the skull base.

The endonasal approach can be used to access deep lesions; however, we would not endorse this approach as the bulk of the tumor lies behind the pituitary gland, and transposition of the gland with its associated risks make this maneuver undesirable. Furthermore, when large vessels are involved, or when tumors extend laterally into the middle cranial fossa, dissection become difficult, dangerous, or even impossible to perform via the endonasal approach. Lastly, a poorly aerated sphenoid sinus significantly hinders the efficiency of this approach.

Approach of Choice

As we are faced with a potentially bloody meningioma with multiple vessel and cranial nerves either entrapped or displaced, we believed that a supratentorial approach would be best suited for dissection of these critical structures out of harm's way. Furthermore, in our experience, the balance of risks to benefits associated with performing two common keyhole approaches is more favorable than with a single, large, less commonly employed, and more invasive operation such as the presigmoid–transpetrosal approach. Our surgical plan was therefore set for a two-stage approach, starting with an eyebrow incision/keyhole craniotomy for a subfrontal approach using standard microsurgical techniques and endoscopic assistance, followed by a retrosigmoid craniotomy for the clival portion if necessary.

Description of the Technique

An eyebrow (transciliary) supraorbital subfrontal approach gives the surgeon excellent access to the ipsilateral anterior cranial fossa (ACF), the medial contralateral ACF, the planum and tuberculum sphenoidale, the suprasellar region, the medial posterior middle cranial fossa, the posterior clinoid region, and to the superior one-third of the clivus (**Fig. 12.9**). Of note, the floor of the anterior middle fossa tucked under the sphenoid ridge is *not* generally accessible through the eyebrow approach, and therefore, tumors which extend to the middle fossa floor require either a subtemporal or a pterional approach.

Our technique to perform an eyebrow approach is effective from both a surgical access and cosmetic standpoint. The patient was positioned supine with the head in gentle extension and contralateral rotation to let the ipsilateral frontal lobe elevate. A temporary tarsorrhaphy was performed, and a skin incision within the eyebrow was made just medial to the supraorbital notch to the end of the hair-bearing skin, and as superior as possible to assist in superior retraction of the scalp (**Fig. 12.10a**). The frontalis muscle was divided, and the subgaleal space was undermined to allow a sizable pericranial cuff to be cut and turned inferiorly (**Fig. 12.10b**). A single burr hole just under the anterior attachment of the temporalis muscle was drilled, and a craniotomy flap was created as flush with the orbital roof as possible (**Fig. 12.10c**), as superior as possible, and carried medially to the supraorbital notch.

The dura was dissected away from the ACF floor, and the inner portion of the calvarium and bony protuberances of the orbital roof were drilled flat extradurally (**Fig. 12.10d**). The dura was then opened in a C shape and hinged inferiorly. A cottonoid was placed over the inferior surface of the frontal lobe, and a careful approach to the suprasellar CSF cistern was performed. CSF was patiently drained by opening the suprasellar cistern, and frontal lobe relaxation was obtained. This approach provided excellent access to the inferior frontal lobe, the suprasellar region

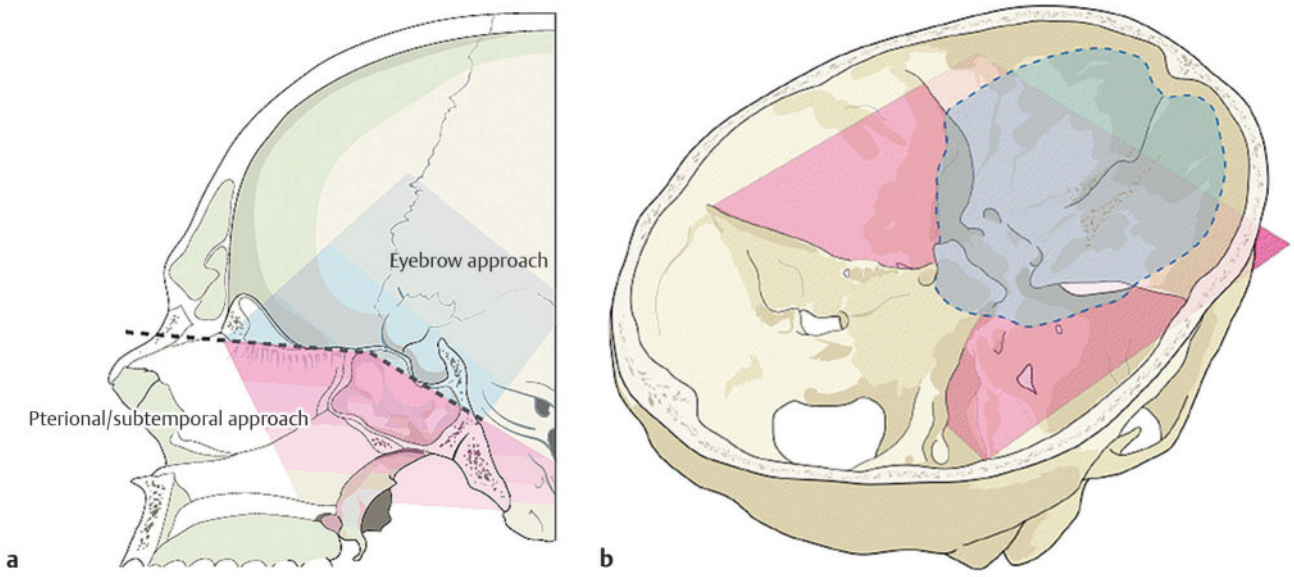


Fig. 12.9 Schematics demonstrating the boundaries of the eyebrow approach. (a) In the sagittal plane, and **(b)** from a superior viewpoint. The eyebrow approach boundaries are indicated in blue, while areas in purple should be accessed via the subtemporal or pterional approach. (Reproduced from Teo C, Sughrue M, eds. Principles and Practice of Keyhole Brain Surgery. 1st ed. Thieme; 2015.)

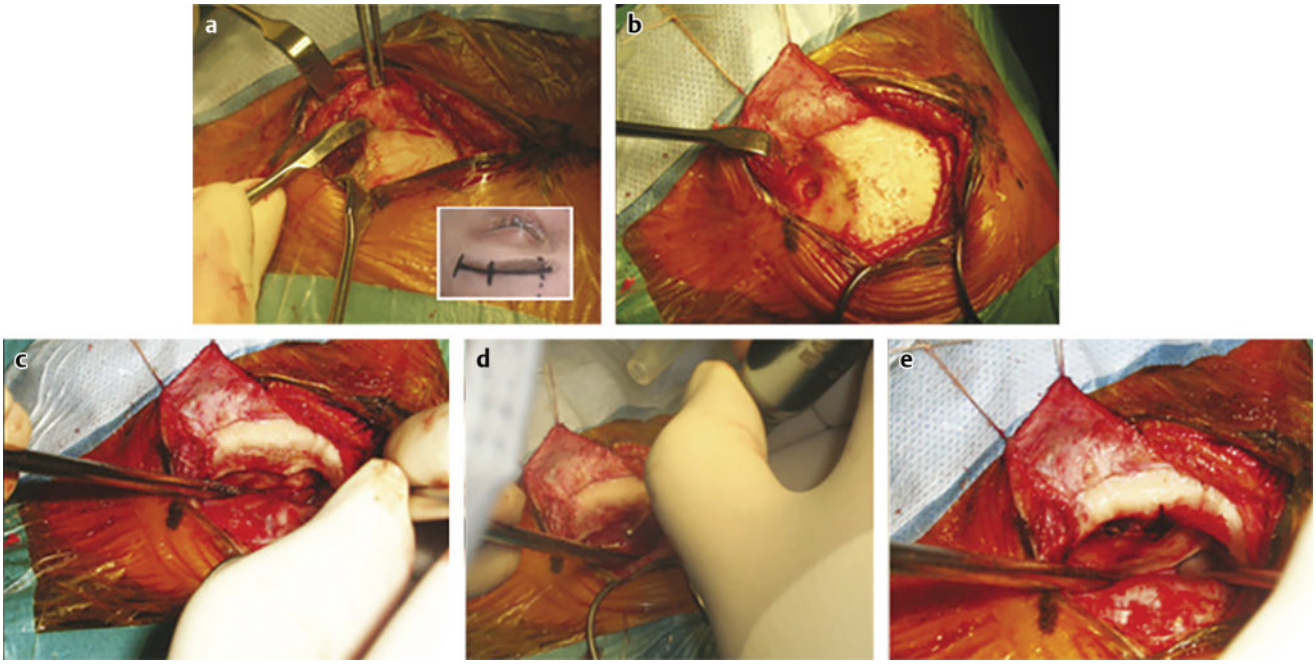


Fig. 12.10 Steps of the eyebrow approach. (a) Inset: the incision is made in the brow and extends from slightly medial to the supraorbital notch to the edge of the brow laterally, and when the eyebrow is thick the incision is preferably closer to the upper border. The soft tissue work aims to expose the lateral orbital rim. In this image, the cut edge of the frontalis muscle is seen, as is the pericranial flap which is cut underneath the frontal side of the skin incision and elevated anteriorly with a small corner of the temporalis muscle. The soft tissue work needs to expose the lateral orbit until the frontozygomatic suture is identified, and until the edge of the superior orbital rim is palpable; **(b)** a single burr hole is made beneath the temporalis at the keyhole. **(c)** Following the creation of a bone flap which is as close to the frontal floor as possible, frontal dura is stripped off the orbital roof in preparation to flatten the orbital roof prominences. **(d)** These prominences should be drilled until completely flush with the sphenoid wing as this will provide more room, and make cerebrospinal fluid access easier during the initial approach; **(e)** demonstrates an adequately flattened orbital roof. Note that the inner table of the frontal bone is also drilled away to improve the line-of-sight to the skull base. (Reproduced from Teo C, Sughrue M, eds. Principles and Practice of Keyhole Brain Surgery. 1st ed. Thieme; 2015.)

including the pituitary stalk and optic apparatus, the hypothalamus and third ventricle, and posteriorly back to access the interpeduncular cistern. In fact, the deep and medial aspects of the sylvian fissure were accessible and opened. With the use of the 30-degree angled endoscope, the approach was expanded to access the upper third of the retroclival region (**Fig. 12.11**).

Using the technique described above and demonstrated in **Fig. 12.12**, the majority of the tumor was removed successfully in the first stage without complications.

■ Aftercare

A postoperative MRI revealed only a small amount of residual tumor near the posterior wall of the cavernous sinus, and as such, the decision was made to postpone the planned second-stage retrosigmoid operation, and follow the tumor with serial imaging (**Fig. 12.13**). The residual disease in the posterior fossa has remained stable for the last 6 years and has not required either the second stage of surgery or any adjuvant treatment.

■ Commentary

The eyebrow approach is extremely versatile. A large frontal sinus, once thought to be a relative contraindication, should be closed with bone wax in a watertight fashion if breached and not cranialized or exenterated as was once recommended. In our experience, mucocele formation is very rare. The opticocarotid

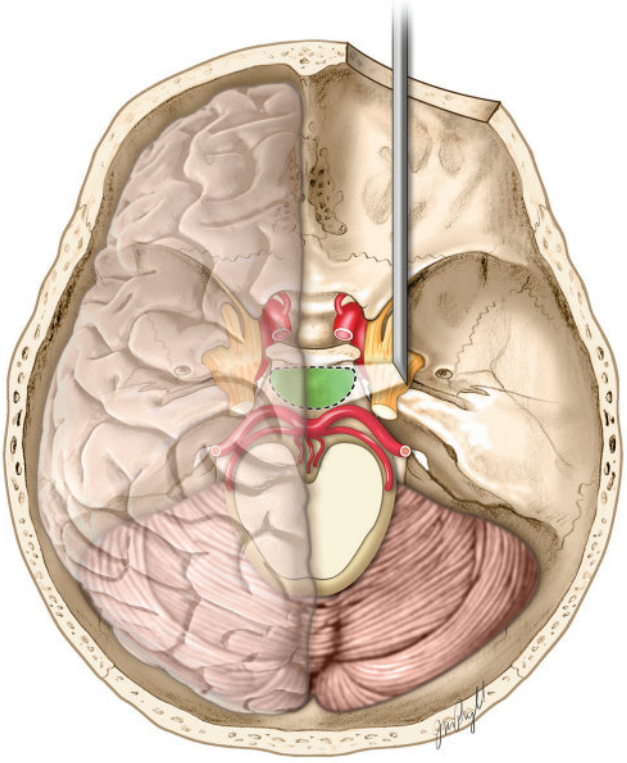


Fig. 12.11 Viewing the retroclival area with an angled-endoscope. Inserted in the carotid–oculomotor triangle, the angled endoscope provides a good view of the posterior clinoid process. It can be angled downward to visualize the retroclival space.

window gives the best access to the posterior clinoid region and may appear insufficient at first glance (**Fig. 12.12a**). Gentle and precise dissection will increase the size of this corridor and allow safe access to the posterior clinoid. It should be noted that while the eyebrow approach can give access to the medial and posterior parts of the middle fossa, in our experience the floor of the anterior middle fossa has been historically inaccessible via this approach. In the original case presented by our colleagues, **Fig. 12.1d** appears to show in the coronal plane the tumor does not extend down to the floor.

Watertight closure of the dura is more important here than with other cranial approaches. Extradural CSF transgression results in a swollen periorbital area, eyelid closure, delayed hospital discharge, and an unhappy patient. Once the dura is closed, the dural incision is covered with Gelfoam, Surgicel, and fibrin glue if there is discernible CSF leakage. Poor replacement of the bone flap will result in long-term cosmetic consequences. The gap between the flap and the edges of the defect should be inferior (underneath the eyebrow), not in the more visible area superiorly. Replacement of the pericranial flap also contributes to a good cosmetic result. Precise and gentle soft tissue manipulation while closing the eyebrow incision is mandatory. A thick scar from poor technique will result in an obvious cosmetic deformity, while

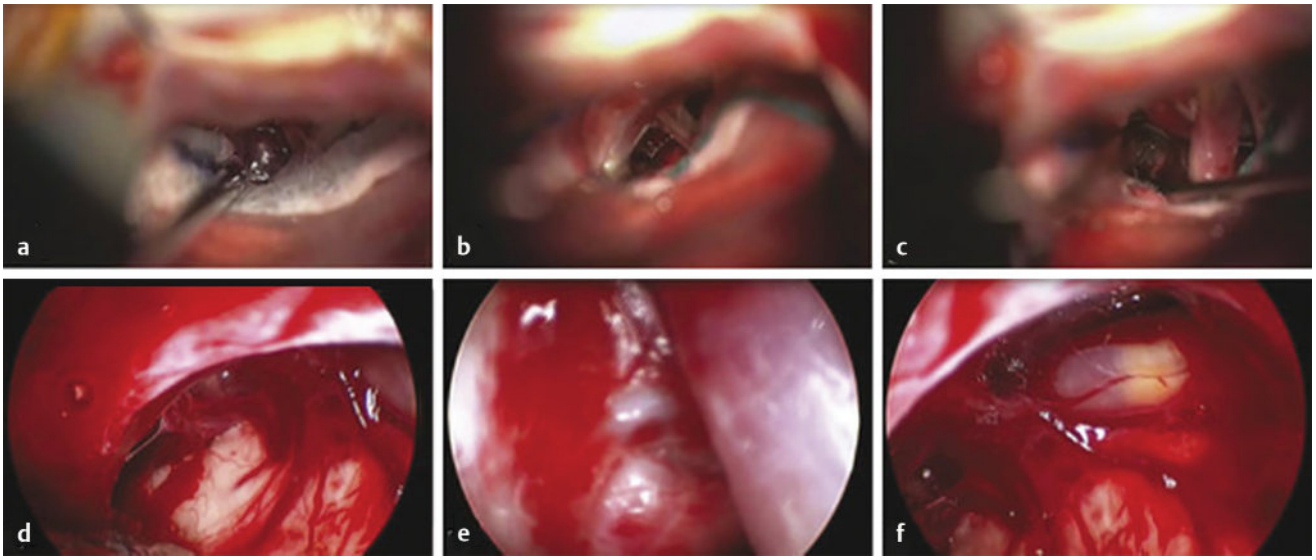


Fig. 12.12 Resection of a posterior clinoidal meningioma via the eyebrow keyhole approach. (a) Arachnoidal dissection is used to identify the opticocarotid and carotid–oculomotor triangles. **(b)** While tumor work through the opticocarotid triangle is usually preferable, the posterior clinoid is best viewed through the carotid-oculomotor triangle **(c)** which is useful in these cases. **(d)** view of the retrodorsal space with a 30-degree endoscope, angled downward to obtain aggressive resection of the tumor behind the dorsum sellae. The top of the dorsum sellae is at the top of the screen, and the prepontine space can be seen behind this. **(e)** View of the posterior clinoid as the endoscope is inserted into the carotid–oculomotor triangle. **(f)** The endoscope is angled medially to view the prepontine space and the back of the posterior clinoid, to ensure complete resection. (Reproduced from Teo C, Sughrue M, eds. Principles and Practice of Keyhole Brain Surgery. 1st ed. Thieme; 2015.)

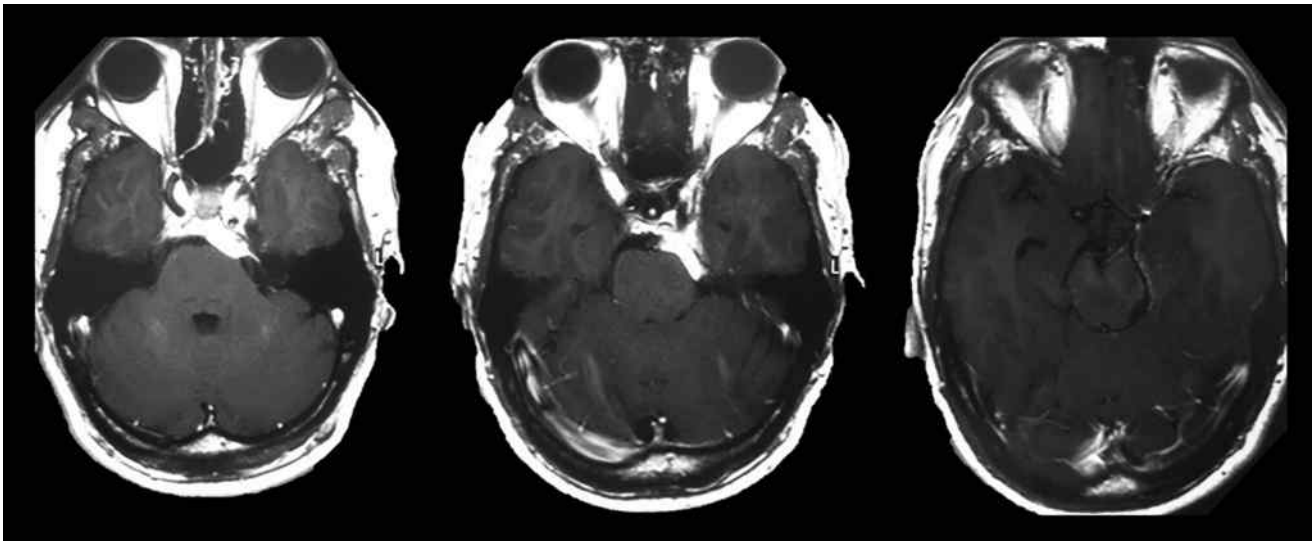
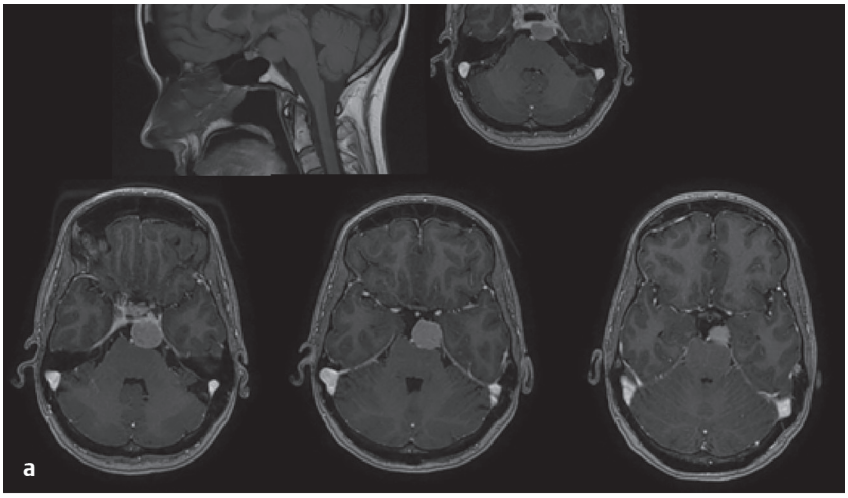


Fig. 12.13 Postoperative MRI. Axial images showing the extent of resection via the eyebrow approach. The residual tumor has been stable for 6 years as of the latest follow-up.

diligent attention to detail can produce a fantastic cosmetic result (**Fig. 12.14**).

Had a second stage been required in our patient, the retrosigmoid approach would have been the best choice. It provides excellent access to the middle third of the clivus, an area not accessible through the eyebrow approach, and can reach tumors with further inferior extensions into the posterior fossa (**Fig. 12.15**). For this procedure, the patient is positioned supine maximally slid to the ipsilateral edge of the bed, with the head slightly elevated above the shoulder and turned maximally to the contralateral side. Use of the frameless navigation to identify the transverse–sigmoid junction, followed by a 2-cm craniectomy, gives excellent access to the anterior and superior surfaces of the cerebellum en route to the retroclival region. It is essential to make the cranial opening as anterior as possible, as a suboptimal opening may necessitate more retraction of the cerebellum. To ensure the bony opening is ideally located, the posterior margin of the sigmoid sinus should be seen before making the durotomy. The posterior bone edge of the craniotomy should not be too far forward, as the bony edge can obstruct the view to deeper, more anterior pathology. Again,

Fig. 12.14 Postoperative cosmetic result of the eyebrow.



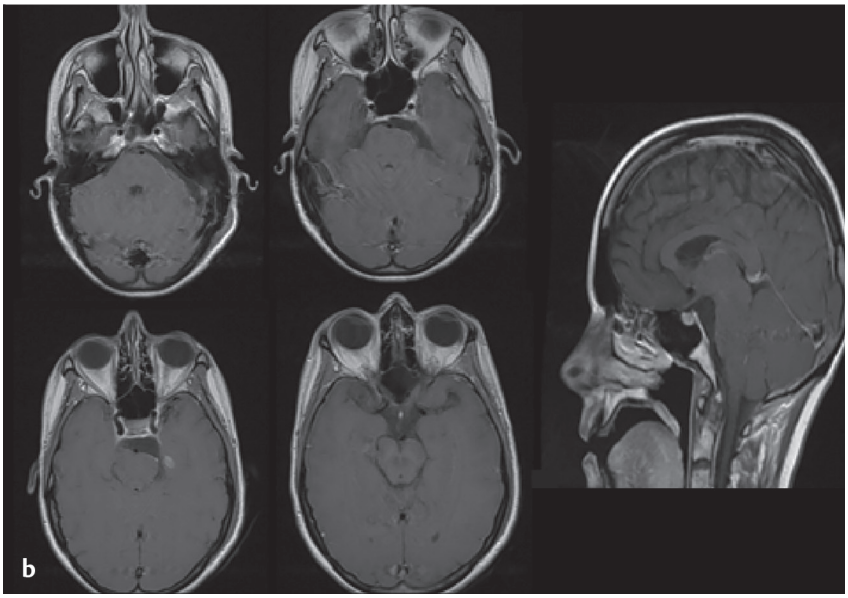


Fig. 12.15 Posterior clinoidal meningioma removed through a retrosigmoid approach. **(a)** Preoperative imaging demonstrating a meningioma of the posterior clinoid. There is no ideal, simple approach to deal with this tumor, the goal being a complete removal for this young patient. The challenge with this tumor is that it lies partially below the eyebrow plane, but above the region that can be comfortably reached with a subtemporal approach. A minipterional approach could reach much of this tumor, but it would be challenging to reach down to the cerebellopontine angle without significant temporal lobe retraction. Ultimately, the retrosigmoid approach as chosen, as it is generally easier to work upward above the tentorium than back on oneself as would be required in other approaches. The patient was prepared for the possibility that two approaches might be needed. **(b)** Postoperative images demonstrating complete removal. (Reproduced from Teo C, Sughrue M, eds. Principles and Practice of Keyhole Brain Surgery. 1st ed. Thieme; 2015.)

CSF cisterns should be opened early and CSF patiently drained to afford necessary brain relaxation to navigate between the crossing cranial nerves and posterior circulation arteries. The vein of Dandy (superior petrosal vein) should be identified early and sacrificed if necessary. Injudicious retraction of the cerebellar hemisphere may result in tearing of this vein and bleeding that is difficult to identify if the cisterns have yet to be opened.

In both the eyebrow and retrosigmoid approaches, once the tumor is encountered, there are other factors that can add to the difficulty of achieving a maximally therapeutic resection of a skull base meningioma. The in situ resection can be impacted by the amount of arterial and neural encasement, and the consistency of the tumor. Efficient use of time and energy should be emphasized, and both the dissection and resection processes should be approached in a systematic way. Ideally, the tumor is devascularized at the base firstly, followed by identification and preservation of the involved nerves and vessels, and then piecemeal removal of the tumor.

■ Conclusion

Tumors arising from the posterior clinoid are typically difficult to remove for reasons that have been thoroughly discussed. We consider surgical cases such as these to be quite important academic and patient care experiences, as they prompt detailed consideration of multiple anatomical and surgical nuances. Coupled with collaborative discussion with colleagues who have their own experiences to share, a surgeon's honest introspection about what is comfortable, safe, and achievable in their hands can make even difficult tumors manageable.

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Part V

Tumors Around the Clivus

13 Petroclival *165*

14 Spheno-Caverno-Petroclival *181*

15 Petroclival Fissure *192*

16 Foramen Magnum *204*

17 Craniovertebral Junction *214*

13 Petroclival

Moujahed Labidi, Kentaro Watanabe, Shunya Hanakita, and Sébastien C. Froelich

Keywords: combined transpetrosal approach, tentorium resection, mobilization of sinus junction, basilar artery

■ Case Presentation

A 42-year-old woman presented for neurosurgical consultation with a 1-year history of headaches, tinnitus, and vertigo. In the few months before the consultation, she also started having episodic, shock-like, left-sided facial pain. Her medical history was otherwise unremarkable. On neurological examination, she was found to have paresthesias on the left side of her face, but the rest of the cranial nerves were normal. The magnetic resonance imaging (MRI) revealed a homogeneously enhancing petroclival lesion, measuring 26 × 33 × 29 mm and causing significant mass effect on the brainstem (**Fig. 13.1**). There was no evidence of hydrocephalus.

Questions

1. How can you explain the patient's facial pain?
2. Other than surgery, are there any other management options?
3. If surgery is considered, how would you conduct the preoperative evaluation?

■ Diagnosis and Treatment

The imaging characteristics of the lesion, including its location medial to the internal acoustic meatus, the enhancement pattern, and the presence of a dural tail, are consistent with a petroclival meningioma. The differential diagnosis includes metastatic lesions, trigeminal schwannoma, and other rare extra-axial tumors such as hemangiopericytoma and chordoma. More so than meningiomas existing elsewhere in the skull base, those in the petroclival region should be treated conservatively whenever possible. A "watchful waiting" strategy and conservative symptom management are usually the first options. In the case of this young woman, various medications have been tried without success, including neuromodulating agents such as gabapentin and pregabalin.

If a patient's symptoms are mostly attributable to hydrocephalus caused by obstruction of the cerebrospinal fluid (CSF) pathway by the petroclival meningioma, ventriculoperitoneal shunting can be a good alternative, particularly in an elderly patient. A shunt can delay or avoid surgery altogether, as the development of hydrocephalus does not necessarily predict continued tumor growth. Radiation therapy is not a good option as stand-alone treatment for most petroclival meningiomas. This is especially true when there is significant brainstem compression, as radiation rarely leads to significant size reduction in meningiomas. The only role of radiation, in the form of radiosurgery, is for small tumors, or as adjuvant treatment of residual disease after surgery.

Returning to our specific patient, a follow-up MRI was performed at 3 months after initial encounter, and although there was only slight volumetric increase of the tumor, her symptoms had worsened. Considering her progressive symptoms, young age, and the tumor's relatively large size, surgical intervention

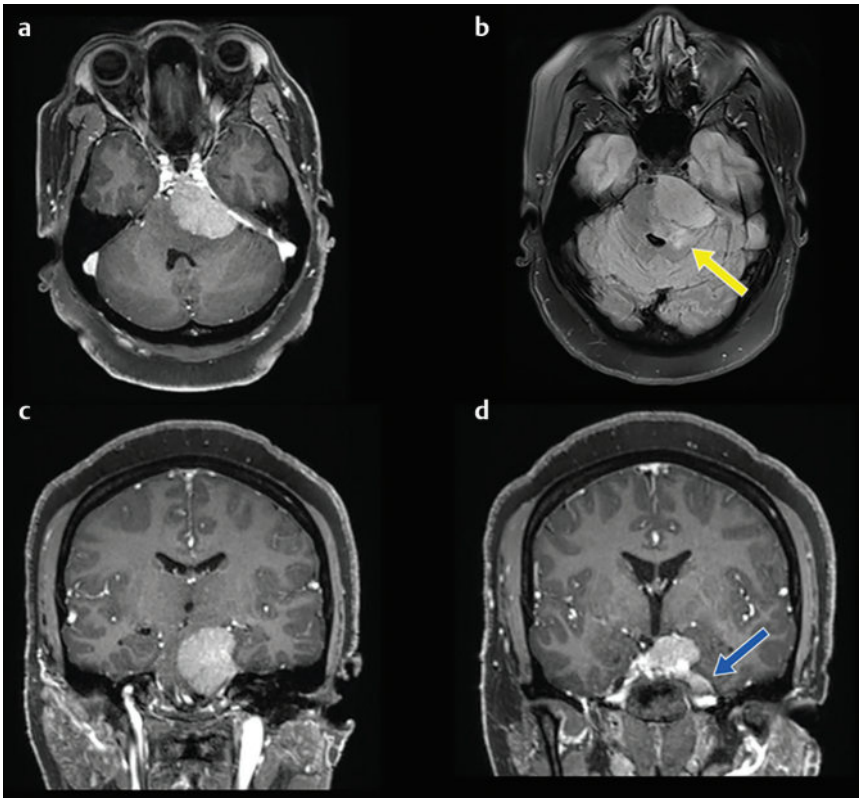


Fig. 13.1 Preoperative images. (a) Contrast-enhanced T1-weighted axial image. **(b)** FLAIR axial image. The *yellow arrow* indicates brainstem edema. **(c)** Contrast-enhanced T1-weighted coronal image. **(d)** Contrast-enhanced T1-weighted coronal image. The *blue arrow* indicates Meckel's cave extension.

was recommended with the goal of brainstem decompression and maximal resection without iatrogenic injury.

■ Anatomical and Therapeutic Considerations

Before undertaking surgery for a petroclival meningioma, a complete preoperative work-up should include assessment of the cardiopulmonary and thromboembolic perioperative risk. Cranial nerve evaluation should include endoscopic assessment of the vocal cords and swallowing function. Preoperative deficits of lower cranial nerves often require a tracheostomy and/or feeding tube (i.e., gastrojejunostomy) in the postoperative period, and patients should be forewarned about these issues. An audiogram is necessary to determine if hearing is serviceable on the side of the meningioma. The MRI should include volumetric *constructive interference in steady state* (CISS), *fast imaging employing steady-state acquisition* (FIESTA), or similar sequences to assess cranial nerves and their spatial relationship with the tumor. A computed tomography (CT) scan is helpful in the assessment of the bony anatomy, including tumor calcifications, pneumatization of the mastoid bone and petrous apex, dehiscence of the carotid canal, high position of the jugular bulb, position of the cochlea and superior semicircular canal, and geniculate ganglion dehiscence if anterior petrosal approach is considered. Along with MR venogram, a cerebral angiogram and venogram can be useful in identifying the major feeding vessels of the meningioma and pattern of venous drainage (occlusion of superior petrosal sinus (SPS), location and drainage of the vein of Labbé, sigmoid sinus dominance, etc.). Moreover, preoperative embolization of difficult-to-reach arterial feeders, such as the ascending pharyngeal artery, and branches of the meningohypophyseal and inferolateral trunks of the internal carotid artery (ICA), can be extremely helpful.

Taking together all the results from our preoperative evaluation, we were faced with a large petroclival tumor that invaded into Meckel's cave with significant extension medial and inferior to the internal auditory meatus (IAM). The patient's hearing was intact and the mastoid was adequately pneumatized. There was edema in the superior cerebellar peduncle and the preoperative angiogram demonstrated two significant arterial feeders. The first arose from the accessory middle meningeal artery (branch of the internal maxillary artery) and the second from the posterior meningeal branch of the ascending pharyngeal artery (**Fig. 13.2**).

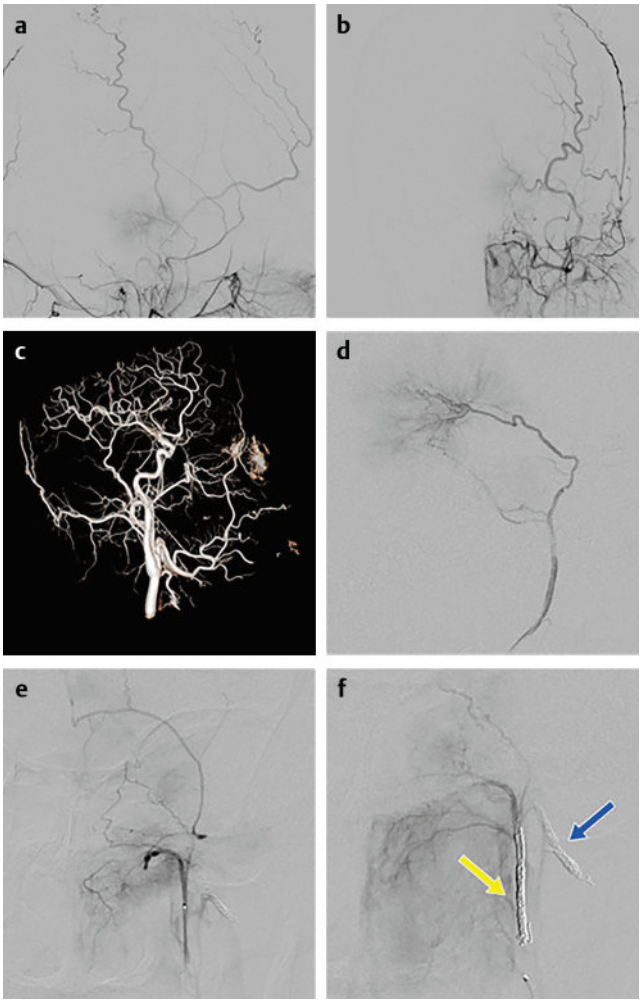


Fig. 13.2 Preoperative angiogram. (a) Lateral and **(b)** anteroposterior projection views of a digital subtraction angiogram (DSA) following selective injection of the left external carotid artery. The tumor blush associated with the meningioma is readily identified. The main arterial feeders of the tumor were two posterior branches of the accessory meningeal artery and the meningeal trunk of the ascending pharyngeal artery. **(c)** 3D reconstruction of a DSA acquisition following internal carotid artery injection. **(d)** Lateral projection views demonstrating elective catheterization of the accessory meningeal artery right before its division into its two branches. Injectable microparticles (300–500 µm) were used to fill the distal feeders close to the tumor, followed by embolization of the trunk of the accessory meningeal artery with injectable microcoils. **(e)** The meningeal trunk arising from the ascending pharyngeal artery was also embolized, as shown in this anteroposterior projection view. **(f)** Final lateral projection view showing coils in both the accessory meningeal artery (*blue arrow*) and meningeal trunk of the ascending pharyngeal artery (*yellow arrow*).

Options for Approach

Several surgical approaches can be used to resect petroclival meningiomas, including the retrosigmoid craniotomy, anterior transpetrosal approach, combined transpetrosal approach, or even the extended endoscopic endonasal approach (EEA).

The retrosigmoid craniotomy is a quick approach into the posterior fossa familiar to all neurosurgeons. It can provide sufficient exposure to small-to-medium-sized tumors and is associated with less risk to the venous structures around the petrous bone. However, the trajectory to the tumor is through smaller windows between the cranial nerves, with only one line-of-sight. For large, calcified, and/or highly vascular tumors, significant retraction on the cerebellum would be required to provide sufficient exposure to the tumor, and even with this, access would still be very limited to the petroclival dura, Meckel's cave, and the tumor interface with the brainstem and vessels, medial to the cranial nerves.

With the refinement of skill and equipment, the EEA has become a viable, "minimally invasive" approach to resect petroclival meningiomas. However, most cases that are suitable for this technique are "clival" rather than "petroclival" lesions. The

advantages of the EEA are that it provides wide and early access to the clival base of the meningioma and thus early devascularization of the medial arterial feeders. It also obviates the need for temporal or cerebellar retraction. In cases of "clival" meningiomas, a wide midline view of the posterior fossa is obtained and the surgical trajectory does not intersect with the paths of the cranial nerves. However, in lesions with lateral extensions (true "petroclival"), it may be difficult to obtain satisfactory tumor resection without mobilization of the ICA. Additionally, even with adequate reconstruction techniques using vascularized flaps, the EEA is still associated with a significant risk of postoperative CSF leak and meningitis.

To begin consideration of approaches through the petrous bone, the anterior petrosal approach as a stand-alone option is quick, and provides sufficient access medial to the acoustic–facial bundle and parts of the meningioma involving the cavernous sinus and Meckel's cave. It allows for early devascularization of the arterial feeders arising from the inferolateral trunk and internal maxillary artery. However, the surgical corridor is both narrow and deep, and the reach is limited inferiorly and posteriorly to the level of the IAM. It is ideal for petroclival–tentorial meningiomas with significant brainstem mass effect that pushes the trigeminal nerve laterally, and extensions into the Meckel's cave and above the free edge of the tentorium.

Approach of Choice

In this case, the combined transpetrosal approach, coupling an anterior with a posterior petrosectomy, was chosen because it affords early devascularization at the tumor base and wide exposure of the tumor at its interface with the cranial nerves and brainstem. Additionally, the combined transpetrosal approach allows resection of tumor in Meckel's cave and on the lateral wall of the cavernous sinus.

Highly modular in nature, a posterior petrosectomy can be tailored to the tumor size and specific location, as well as to the hearing status of the patient (**Fig. 13.3**). A retrolabyrinthine petrosectomy is chosen when hearing is still serviceable, as with our patient. In larger lesions associated with hearing loss, a translabyrinthine approach may be used. A transcochlear approach, with rerouting of the facial nerve, has classically been described for even wider exposure, but we have not found it useful, and it entails an unacceptably high risk of facial nerve injury.

The main advantage of the combined petrosal approach is that it provides multiple lines-of-sight, each of which can be exploited for optimal dissection of the various cranial nerves and major vessels entangled by the tumor, thus reducing the risk of injury. In cases where there is brainstem pial infiltration by tumor, the wider exposure and multiple trajectories provide better visualization of the "carpet" of residual tumor that must be left on the brainstem, again making resection safer. Another major advantage is that minimal or no retraction of the cerebellar hemispheres and temporal lobe is needed to resect the tumor with posterior mobilization of the transverse–sigmoid junction (TSJ).

However, the combined transpetrosal craniotomy is a long and tedious operation that may increase "time-dependent" complications such as decubitus ulcers and thromboembolic events. In fact, some surgeons advocate staging the operation. Related to drilling and operating through the posterior petrosectomy, there is a small risk to the facial nerve in the mastoid

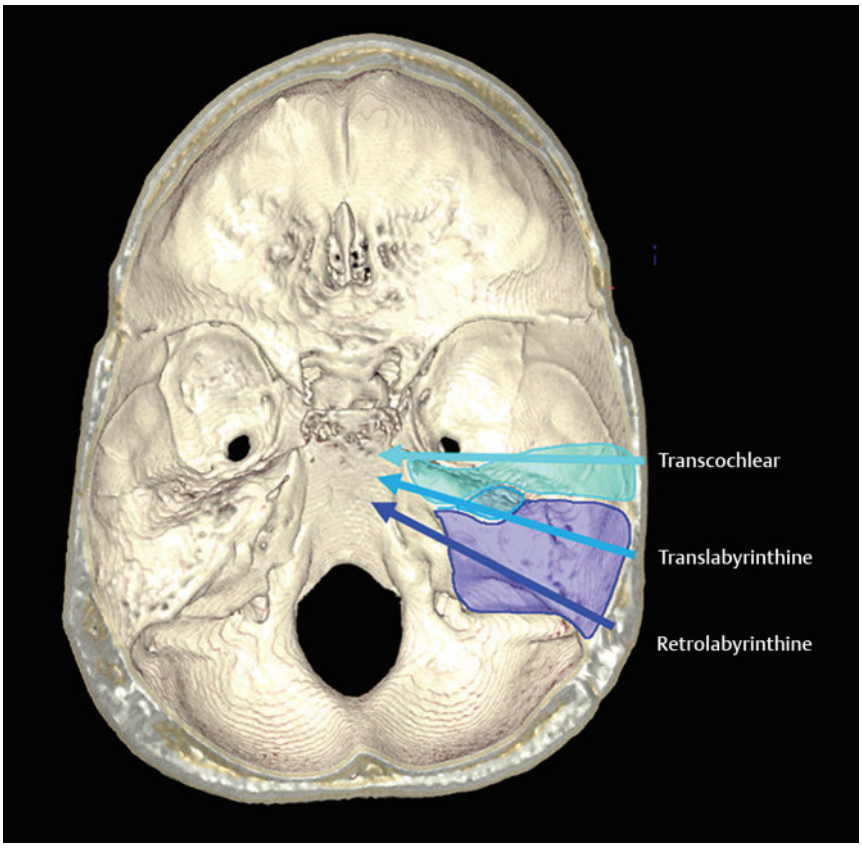


Fig. 13.3 Various forms of the posterior petrosectomy. All of these are "presigmoid," as the surgical trajectory is anterior to the sigmoid sinus. Progressive removal of the petrous ridge places the surgeon more anteriorly, with increasing exposure across the midline. The translabyrinthine approach is used in patients with non-servicable hearing, and the transcochlear approach requires mobilization of the facial nerve which results in facial palsy.

segment, and a larger risk of venous infarction, either secondary to thrombosis/occlusion of the sigmoid sinus or, more importantly, of injury to the vein of Labbé. Detailed knowledge of the patients' specific venous anatomy and their careful manipulation can help reduce venous complications.

Questions

1. What anatomical landmarks can be used to identify the fallopian canal and mastoid segment of the facial nerve during temporal bone drilling?
2. What cranial nerves are most at risk of injury during incision and resection of the tentorium?
3. Can you identify arterial branches that are most often feeders of petroclival meningiomas?
4. What are the surgical implications of brainstem edema and how does it influence your surgical strategy?

■ Description of the Technique

The day before surgery, the patient underwent endovascular treatment. The feeders from the accessory middle meningeal artery, as well as that from the posterior meningeal branch of the ascending pharyngeal artery, were successfully eliminated with injectable microcoils (**Fig. 13.2f**).

The following day, the patient was taken to the operating room. She was positioned in the supine position with a shoulder roll placed beneath the left shoulder. The head, fixed in a three-pin skull clamp, was then rotated 70 to 80 degrees to the right side (10–20 degrees from the horizontal plane) and the head was elevated above the level of the heart. This, in addition to a slight reverse Trendelenburg positioning of the surgical table, ensured good venous outflow to lessen intraoperative bleeding. The vertex was then tilted downward, a maneuver that allowed gravity-assisted "retraction" of the temporal lobe. The abdomen was prepared for harvesting a fat graft to repair the dural defect and fill the mastoid (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral and lateral

Craniotomy: temporal/retrosigmoid (L-shaped)

Modification: anterior + posterior petrosectomies, resection of tentorium, mobilization of TSJ

Navigation with both MRI (T2-weighted or CISS/FIESTA and T1-weighted with gadolinium) and CT imaging fusion, as well as monitoring of motor evoked potentials and electromyography of cranial nerves III (medial rectus muscle), V (masseter muscle), VI (lateral rectus muscle), VII (orbicularis oculi and oris muscles), IX (pharyngeal elevator muscle), X (vocal cord), and XI (trapezius muscle), were utilized.

Incision and Craniotomy

A "C-shaped" curvilinear incision was made, starting from the tip of the mastoid, going posteriorly 2 cm behind the approximate location of the TSJ and encircling the approximate location of the temporalis muscle. The incision should be anterior enough to allow exposure of the pterional keyhole. An "L-shaped" craniotomy was then performed, combining a temporal and a retrosigmoid craniotomy, with the junction of the two straddling the transverse sinus. Two burr holes were made above the transverse sinus and two others below. Connecting these burr holes and crossing the transverse sinus was performed first with the cutting and then the diamond burr to safeguard the sinus, which sometimes indents the occipital bone. The craniotomy was positioned as low on the floor of the middle fossa as possible. The retrosigmoid dura was exposed approximately 2 cm behind the level of the sigmoid sinus to allow for posterior mobilization of the TSJ (see **Operative Setup**).

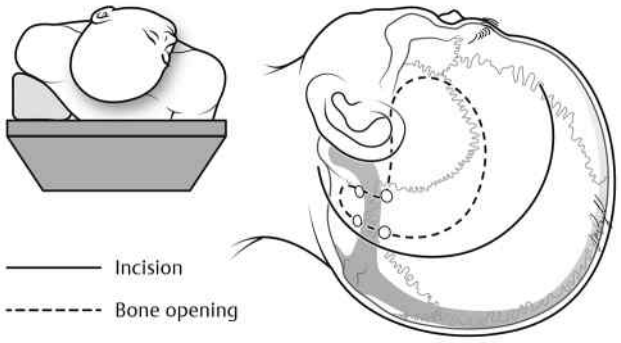
Operative Setup

Position: supine, head rotated 80 degrees, elevated with vertex down

Incision: "C-shaped" from mastoid tip, superiorly and then anteriorly

Bone Opening: "L-shaped," temporal + retrosigmoid

Durotomy: presigmoid + subtemporal above transverse sinus



Petrosectomy

A retrolabyrinthine mastoidectomy was then performed (**Video 13.1**). The outer landmarks defining the mastoidectomy form a triangle, with the mastoid tip inferiorly, the asterion posterosuperiorly, and the root of the zygoma anterosuperiorly (**Fig. 13.4a**). A cosmetic mastoidectomy was first performed by removing the outer cortical bone of the mastoid en bloc with a bone chisel. Then, a large cutting burr with continuous irrigation was used to identify the first major landmarks, which included the temporal dura, the sigmoid sinus, and the sinodural angle (**Fig. 13.5a**). After switching to a diamond burr, the next landmark reached was the mastoid antrum, which was widely opened to allow identification of the incus (**Fig. 13.5b**). The short arm of the incus is a very useful landmark to identify the mastoid segment of the facial nerve that runs inferior to the lateral (horizontal) semicircular canal. Knowing this position allowed the surgical corridor to be safely widened around and below the labyrinth.

At this stage, all three semicircular canals were completely skeletonized and the bone overlying the infralabyrinthine dura was removed (**Fig. 13.4b**). The endolymphatic sac was disconnected from the vestibular aqueduct at the level of the posterior

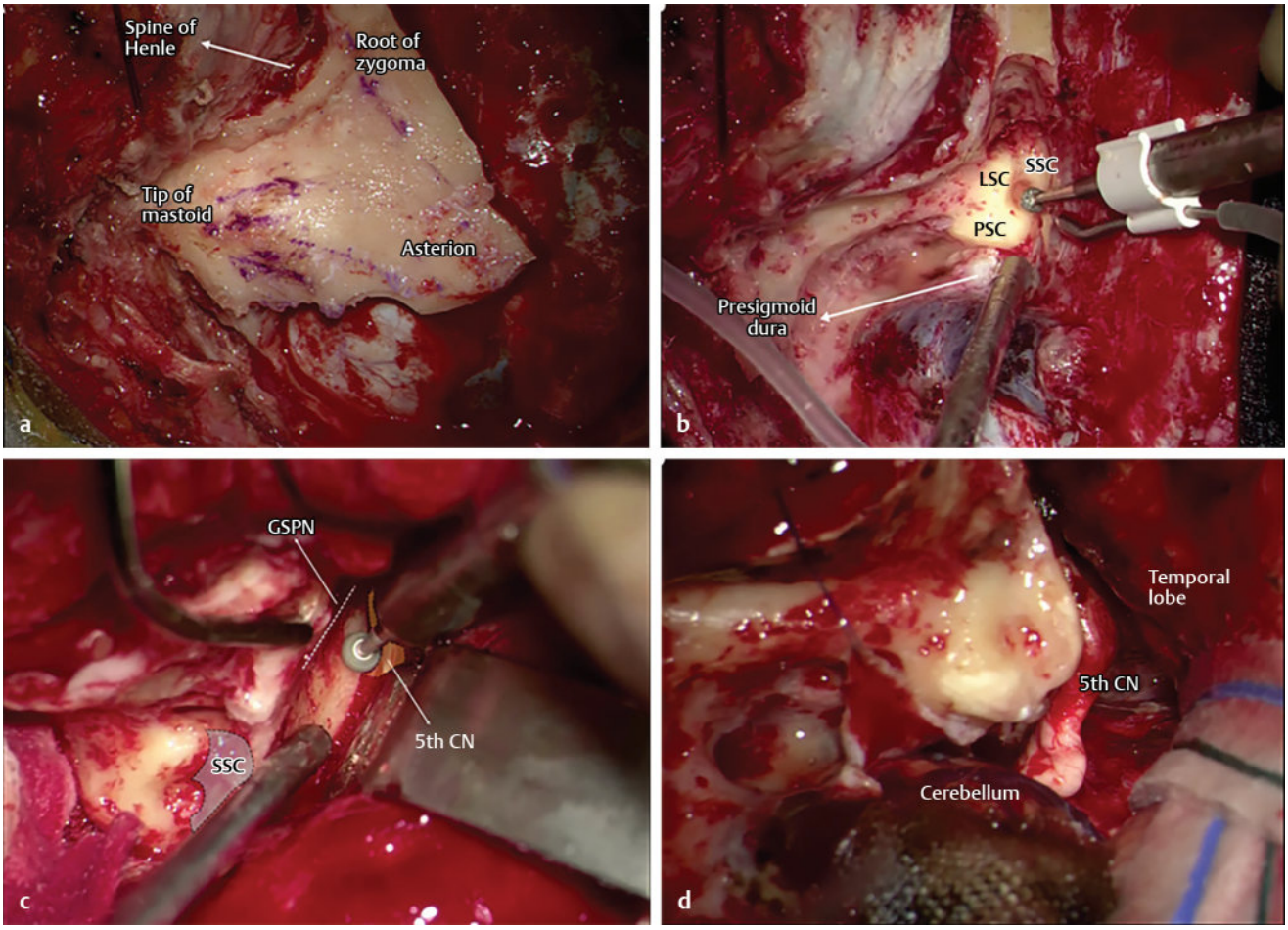


Fig. 13.4 Intraoperative images. (a) The exocranial limits of the mastoidectomy form a triangle; the asterion posterosuperiorly, the root of the zygoma anterosuperiorly, and the tip of the mastoid, inferiorly. The spine of Henle marks the external acoustic meatus. **(b)** Skeletonization of the semicircular canals during retrolabyrinthine mastoidectomy. LSC, lateral semicircular canal; PSC, posterior semicircular canal. SSC, superior semicircular canal; **(c)** An anterior petrosectomy was then performed in Kawase's rhomboid, delimited laterally by the GSPN, posteriorly by the SSC (the arcuate eminence has been drilled down to the SCC), anteriorly by the fifth cranial nerve and medially by the petrous ridge. CN, cranial nerve; GSPN, greater superficial petrosal nerve; SSC, superior semicircular canal. **(d)** Exposure obtained after completing the combined transpetrosal approach.

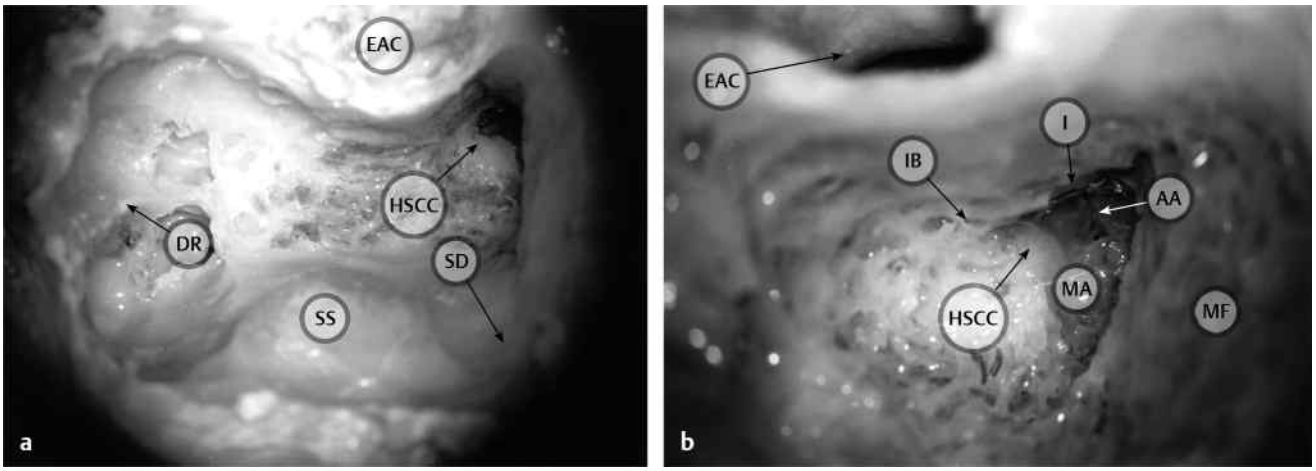


Fig. 13.5 Cadaveric dissection, left-sided mastoidectomy. (a) Removal of posterior (perisigmoid sinus) and inferior (mastoid tip) cells reveals structures such as posterior wall of external auditory canal (EAC), horizontal semicircular canal (HSCC), sigmoid sinus (SS), sinodural angle (SD), digastric ridge (DR). **(b)** By opening the lateral wall of the mastoid antrum (i.e., Koerner's septum) and proceeding anteriorly, the short process of the incus becomes visible. AA, aditus ad antrum; EAC, posterior wall of external auditory canal; HSCC, horizontal semicircular canal; I, short process of incus; IB, incus buttress; MA, mastoid antrum; MF, middle fossa plate. (Reproduced from Adunka O, Buchman C, eds. Otology, Neurotology, and Lateral Skull Base Surgery. An Illustrated Handbook. 1st ed. Thieme; 2010.)

semicircular canal. This allowed further drilling around the labyrinth and posterior mobilization of the sigmoid sinus. The transverse and sigmoid sinuses were uncovered with a large diamond burr, and the remaining thin shell of bone over the sinuses was removed with a small and smooth-tipped rongeur.

An anterior petrosectomy was then performed. The temporal dura was first elevated from the middle fossa from a posterior-to-anterior direction. The middle meningeal artery was sectioned at the foramen spinosum. The greater petrosal nerve was identified, as was the arcuate eminence. The temporal fossa dura was then peeled from V3 and partially from V2 until Meckel's cave came into view. After identification of the landmarks, drilling of the middle fossa rhomboid (Kawase's quadrilateral) was done (**Fig. 13.4c**). The drilling also extended posteriorly above and behind the IAM and under the mandibular division of the trigeminal nerve (V3) in order to maximize exposure of the posterior fossa dura and improve devascularization of the tumor.

Dural and Tentorial Incisions

After completion of the bone work, the posterior fossa (presigmoid) and temporal dura were incised. This step is particularly important in achieving posterior mobilization of the TSJ. A first incision was made parallel and anterior to the sigmoid sinus (the presigmoid incision) (**Fig. 13.6**). Then, a temporal incision was made along the base of the middle fossa extending above the transverse sinus. We usually extend this incision way beyond the TSJ posteriorly, 1.5 cm above the TS to avoid any risk for the temporal veins draining into the TSJ. Incisions were then made above and below the SPS.

Tentorial resection started first by coagulation, ligation, and section of the SPS posteriorly. To preserve venous outflow, this is ideally done just anterior to the junction between superior petrosal veins and the SPS. A posterior incision of the tentorium was then made toward the free edge of the tentorium with direct visualization and protection of the trochlear nerve. The anterior incision of the tentorium should be adapted to the meningioma extension pattern, and for our patient with her large tumor, the Meckel's cave was opened first starting along the lateral aspect of V3 cutting posteriorly, and then an incision was made at the superior margin of the *porus trigeminus* aiming medially and posteriorly through the tentorium. Major feeding vessels encountered during the sectioning of the tentorium were coagulated and cut. In some cases, we have found use for indocyanine green video angiography to tailor the dural and tentorial cuts. This freed piece of tentorium was then removed which opened a large window for tumor debulking.

After this stage, the TSJ can be safely and significantly mobilized posteriorly providing a greatly widened surgical corridor which combined the presigmoid, the retrosigmoid, and subtemporal corridors (**Fig. 13.4d**). Care was taken to ensure patency of the sinuses and of the vein of Labbé during this maneuver with intermittent monitoring with the Doppler probe. The tumor was disconnected from its petrous and clival base to achieve further devascularization. After extensive debulking, the tumor was dissected away from the brainstem and cranial nerves, starting laterally. The trochlear nerve was traced and preserved anatomically. The abducens nerve was identified medial to the tumor and likewise preserved. Special care was taken at the level of the superior cerebellar peduncle, where there was significant edema on the fluid-attenuated inversion recovery sequence. Often, this is a sign of significant adherence between the pial surface and the tumor, and in such a case, it is better to leave a small piece of tumor than to cause brainstem injury. In this particular case, a clear plane was found (**Video 13.1**). Except for a small extension into the cavernous sinus, the tumor was fully resected.

Closure

A well-designed reconstruction is an essential part of the surgical plan. Primary dural closure is preferred, but often impossible after an anterior petrosectomy or combined transpetrosal approach. For our patient, the dura was first reapproximated with sutures, and an onlay graft of pericranium plus temporalis fascia was positioned over the defect and sutured in place. The mastoid antrum was covered with a thin layer of bone wax, fascia, and fibrin glue. The space created by the posterior petrosectomy was filled with autologous fat and fibrin glue. The mastoid cortical bone and the bone flap were replaced and attached with miniplates. The rest of the closure was done in routine fashion.

Surgical Pearls

1. Preoperative cerebral angiography and venogram provide critical information for petroclival meningiomas. When feasible and safe, embolization of arterial feeders can facilitate resection of the tumor and reduce intraoperative blood loss. Most important feeding arteries to petroclival meningiomas are branches arising from the inferolateral trunk and the meningohypophyseal trunk of the ICA. The middle meningeal artery and the ascending pharyngeal artery can also supply these tumors. In cases where there is brainstem edema, there often is blood supply from the pial surface, which cannot be embolized.
2. The surgical corridor in a combined transpetrosal approach is not only through the petrosectomies, as the posterior mobilization of the TSJ can provide wide access to the tumor as well. This maneuver, facilitated by strategically placed dural incisions and sectioning of the tentorium, opens a wide path by combining the presigmoid, the retrosigmoid, and subtemporal corridors.
3. Adequate closure is critical to the overall success of the operation. When a CSF leak occurs, it is usually through the middle ear or through an air cell in the middle fossa that was not adequately closed. These areas can usually be identified on the preoperative CT scan (dehiscent middle ear, zygomatic air cell, large pneumatized mastoid, etc.), and they need to be closed individually with great care. Our technique is to first cover the air cell with a thin layer of bone wax, supplemented by a layer of fascia and fibrin glue. Injury to the external auditory meatus should be avoided as it carries a very high risk of postoperative CSF leak.

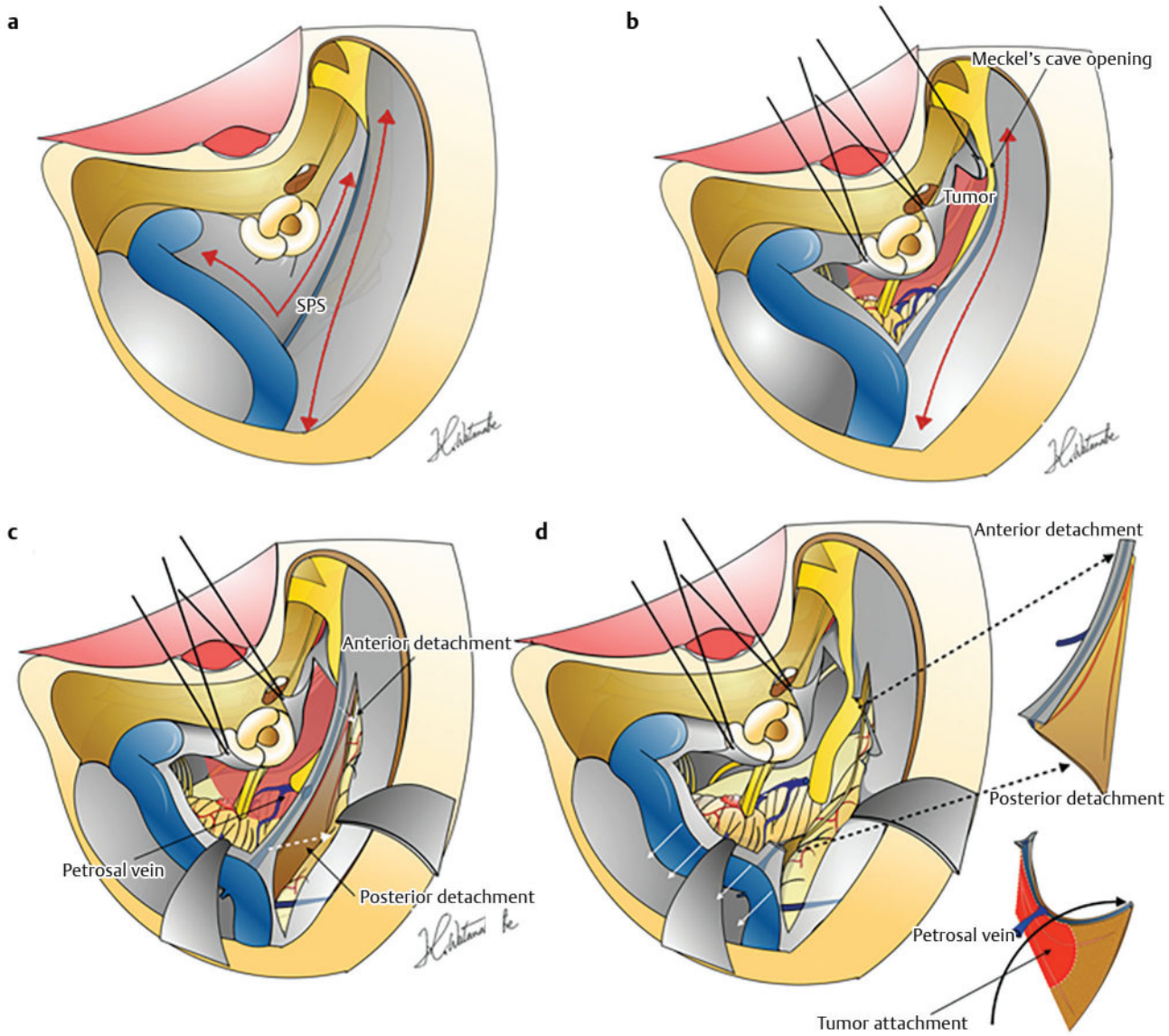


Fig. 13.6 Dural and tentorium incisions. (a) The presigmoid dural incision was made parallel and anterior to the sigmoid sinus (the presigmoid incision) and turned anterior, just below, and following the course of the superior petrosal sinus (SPS). **(b)** After retraction of the presigmoid dural, a temporal incision was done along the base of the middle fossa and extended above the transverse sinus posteriorly. Care was taken to preserve the vein of Labbé and other major venous channels. **(c, d)** Tentorial resection was then done first by coagulation, ligation, and section of the SPS anteriorly. The anterior incision of the tentorium was started at the *porus trigeminus* laterally and directly medially toward the entry of the trochlear nerve in its tentorial tunnel. The posterior incision of the tentorium was made where the superior petrosal vein drains into the SPS. This cut was extended medially toward the free edge of the tentorium while visualizing and protecting the trochlear nerve.

■ Aftercare

All patients should have an MRI in the first 48 hours following tumor surgery to assess the extent of resection and any evidence of complications. Our postoperative MRI showed complete resection of the meningioma, except for a small residual inside the cavernous sinus, and no evidence of retraction injury to either cerebellum or temporal lobe (**Fig. 13.7**). We do not routinely use lumbar drains or antibiotics after surgery, although we do occasionally perform lumbar punctures if patients complain of headaches or show evidence of ventricular dilation on postoperative imaging.

■ Possible Complications and Associated Management

Transient or permanent diplopia due to abducens nerve injury is the most common complications after petroclival meningioma

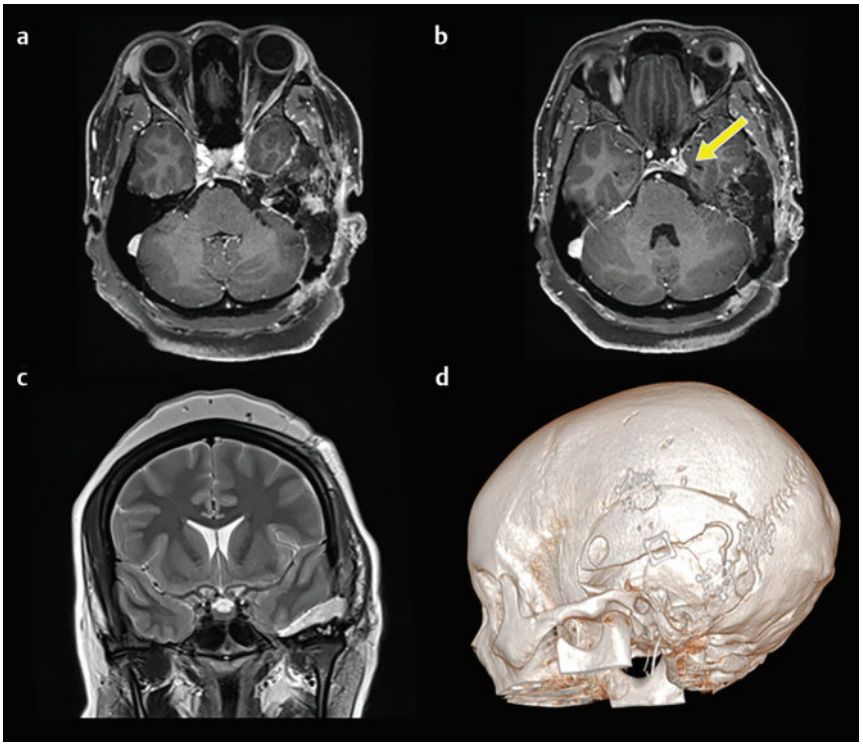


Fig. 13.7 Postoperative images. (a, b) Postoperative axial T1-weighted MRI demonstrating subtotal resection of the meningioma with a small residual in the left cavernous sinus (*yellow arrow*). **(c)** Coronal T2-weighted MRI showing the fat graft atop the floor of the middle fossa and the absence of signal change in the left temporal lobe. **(d)** 3D reconstruction showing the bone flap used and the reconstruction with titanium miniplates.

resection. Trochlear nerve palsy occurs less frequently and has less impact on oculomotility. Other cranial nerves at risk are the trigeminal and the VII/VIII nerve complex. The trigeminal nerve is at risk during dura opening and tentorium resection, as it can be pushed upward and laterally by the tumor immediately behind the dura.

Postoperative CSF leaks and pseudomeningocele are frequent after resection of petroclival meningiomas and range from 2 to 17% and 15 to 28%, respectively. If a CSF leak occurs in the immediate postoperative period, our first line of treatment is usually bed rest and lumbar drainage, unless the leak is deemed to be "high flow." In such cases, immediate surgical reexploration is undertaken. Usually, CSF leakage occurs through the middle ear due to inadequate closure of opened air cells. Pseudomeningocele seldom requires treatment and is usually self-limiting. Brainstem infarction is another complication reported in the literature. The best way to avoid injury to the brainstem is to refrain from overaggressive dissection of meningioma adherent to the pial surface.

Perspective

Walter C. Jean and Timothy R. Deklotz

■ Introduction

The concept of breaking down complex surgery into approach "elements," and picking the ideal elements to construct strategic approaches to complex tumors, was explored in detail in the Introduction of this book. Nowhere is the power of this "building block" concept more clearly showcased than in the demystification of surgeries for petroclival tumors. By separating the petroclival region into anatomical zones, and understanding the approach element most suited to attack each zone, the surgeon only has to assemble the elements to make one coherent operation. Rather than memorizing which massive operation fits which specific kind of petroclival tumor, the decision-making process now borders on being formulaic, and definitely less intimidating.

Meningiomas of the petroclival region straddle the petroclival fissure and vary from each other with differing proportions residing on either side of the fissure: on the petrous side or the clival side. Some surgeons consider only tumors that are medial to the trigeminal nerve as "true" petroclival meningiomas, but whether true or fake, we will consider all meningiomas of the petroclival region in this section. To begin this process, one must first understand the anatomical zones in this region, in which the tumors reside.

■ Zones of the Clivus

Looking at the clivus as a ramp, and following the axis of the vertebral–basilar system, the clivus can be divided into three zones, each of which has an associated approach "element" that is best suited to access it (**Fig. 13.8**, **Fig. 13.9**). Zone 1 extends from dorsum sellae to internal acoustic meatus, and the approach uniquely fitted to attack this zone is the anterior petrosectomy using the lateral corridor. Superiorly located petroclival meningioma often surrounds the proximal posterior cerebral artery and elevates the oculomotor nerve. This approach can provide access to these areas, and by drilling Kawase's quadrilateral, the inferior limit of this approach is to the level of the internal acoustic meatus.

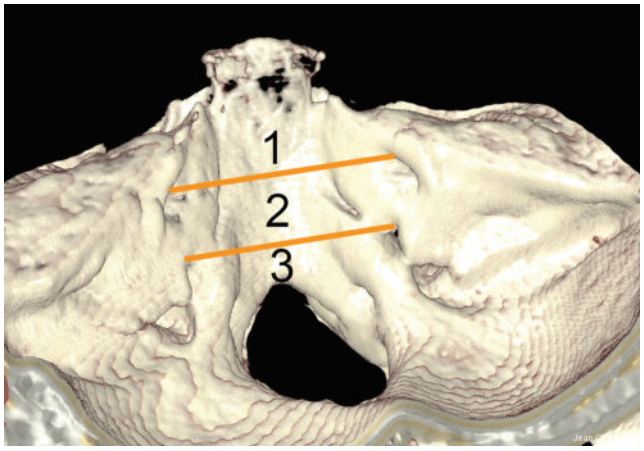


Fig. 13.8 Dorsal view of the petroclival region. Zone 1 of the clivus extends from the dorsum sellae to the line across the two internal acoustic meatus. Zone 2 is between this line and the line across the two jugular foramens. Zone 3 is from the latter line to the foramen magnum.

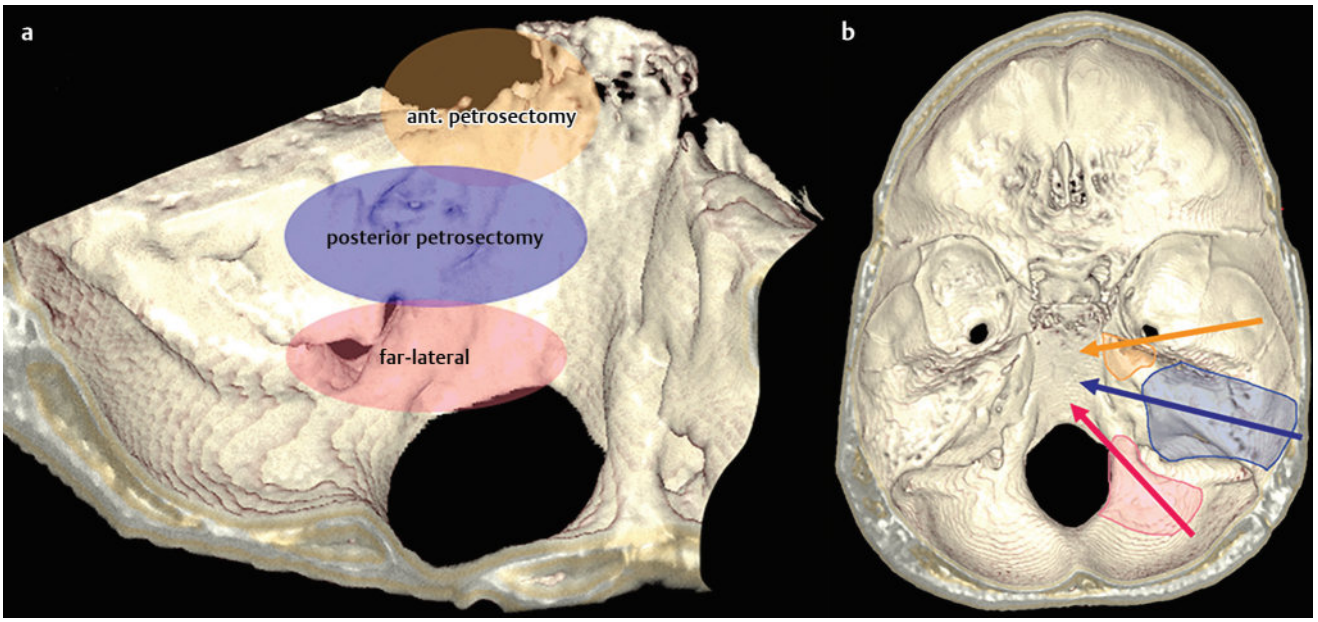


Fig. 13.9 Best-suited approaches to the three zones. (a) Dorsolateral view of the petroclival region. **(b)** Superior view of the same. The anterior petrosectomy is excellent in reaching lesions isolated in zone 1. It can reach superiorly to above the dorsum sellae, and inferiorly to the level of the internal acoustic meatus. The various forms of posterior petrosectomy, including the retrolabyrinthine and translabyrinthine approaches, are fitted to attack zone 2. With incision of the tentorium, their superior reach is almost to the level of the dorsum sellae, and inferiorly they can reach the jugular foramen. The far-lateral approaches, with or without condylectomy, are most useful for zone 3.

Case Presentation 1

A 54-year-old woman presented to clinic with progressively worsening headache for 2 years, multiple endocrinopathies, and deteriorating vision in her left eye. Her tumor extended slightly above the dorsum sellae, and engulfed the posterior cerebral artery and some portions of the middle cerebral artery (**Fig. 13.10a**). Inferiorly, it reached the level of the internal acoustic meatus but not below that. For these reasons, the vast majority of the tumor was resected via the lateral subtemporal corridor, using an anterior petrosectomy to access the petroclival region (**Fig. 13.10b**). An EEA was added to remove the sphenoid sinus component.

Zone 2 of the clivus is between the levels of the acoustic meatus and jugular foramen, and the various forms of the posterior petrosectomies through the posterolateral corridor are the most appropriate approaches to this zone. The basic form of the posterior petrosectomy is retrolabyrinthine (**Fig. 13.3**). By progressively removing more bone, partial or total labyrinthectomy (i.e., translabyrinthine approach) increases the surgical exposure of the posterior petrosectomy, and allows the surgeon more flexibility to access the contralateral side of the clivus. However, partial labyrinthectomy puts hearing at risk, and the translabyrinthine approach is reserved for patients with non-serviceable hearing. The final form of these approaches is the transcochlear approach, now rarely used because it requires posterior mobilization of the facial nerve with its associated morbidities. Although posterior petrosectomies can reach into zone 1, access to the level of the dorsum sellae would require massive retraction forces on the temporal lobe. Inferiorly, visualization is limited below the level of the jugular foramen.

Zone 3 of the clivus is the lowest portion below the line drawn across the jugular foramens. The far-lateral approaches, without or with resection of the occipital condyle (i.e., transcondylar approach) are the best suited approaches for tumors involving this zone. Again, the progressive removal of the occipital condyle puts the origin of the surgeon's line-of-sight more anteriorly, thus augmenting the ability to look across the midline. The superior limit of the approach is at the level of the internal acoustic canal, inferiorly, to C1.

Following the building blocks concept, petroclival meningiomas that extend over two clival zones may require combining approaches for safe removal. The first section of this chapter involved a tumor that straddled zones 1 and 2, and was resected brilliantly by using the "combined transpetrosal" approach. When combining approaches to zones 1 and 2, it is important to remember that the balance and hearing apparatus separate the anterior and posterior petrosectomies, and in the case of zones 2 + 3, it is the sigmoid sinus that separates the presigmoid and far-lateral approaches.

Case Presentation 2

A 37-year-old man presented to the emergency department with progressive difficulty walking for a month. His balance was so poor that he had experienced several falls. His meningioma occupied zones 2 and 3, reaching from slightly above the level of the IAC to the foramen magnum (**Fig. 13.11a**). This tumor was resected via a staged, multidirectional approach, combining a retrolabyrinthine posterior petrosectomy (zone 2) with a far-lateral transcondylar (zone 3) approach through the pre- and retrosigmoid corridors. The operation was staged, with all the bone work done on day 1, and tumor resection on day 2. A near-total resection was achieved with this technique (**Fig. 13.11b**).

■ Endonasal Corridor

With lines drawn across the internal acoustic meatus and jugular foramen on either side, dropping orthogonal lines from these to the midline clivus provides the measurement of the "central clivus depth," which ranges from 4 to 13 mm (**Fig. 13.12**). Tumors attached to the central clival depressions provide serious challenges to lateral or posterolateral approaches as there are "blind spots" behind sharp corners. Ironically, a more "basic" retrosigmoid trajectory, including that gained by posterior mobilization of the TSJ, might visualize these areas better but even so, the large distance-to-target remains daunting (**Fig. 13.13**). However, with the advancement of tools and technique, the EEA through the clivus (i.e., transclival approach) has revolutionized access to these locations.

Applied to petroclival meningiomas, the unique strengths of the endonasal endoscopic transclival approach are early elimination of the tumor blood supply from the clival dura, and relative short working distance when compared to lateral/posterolateral approaches. In this most "standard" form, the approach can access the central clivus from the floor of the sella turcica to the foramen

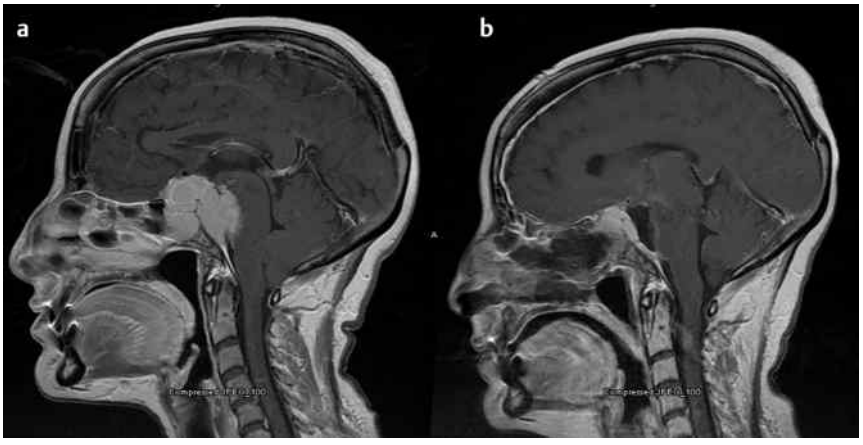


Fig. 13.10 Images of Case 1. This is a superiorly located petroclival meningioma, mostly in zone 1. **(a)** Preoperative MRI showed a tumor that reached slightly above the dorsum sellae down to the level of the internal acoustic canal. On the left, this tumor also engulfed the posterior cerebral artery, small portions of the middle cerebral artery, and the oculomotor nerve. The vast majority of this tumor was removed using the lateral subtemporal corridor with an anterior petrosectomy to reach the petroclival region. **(b)** Postoperative view.

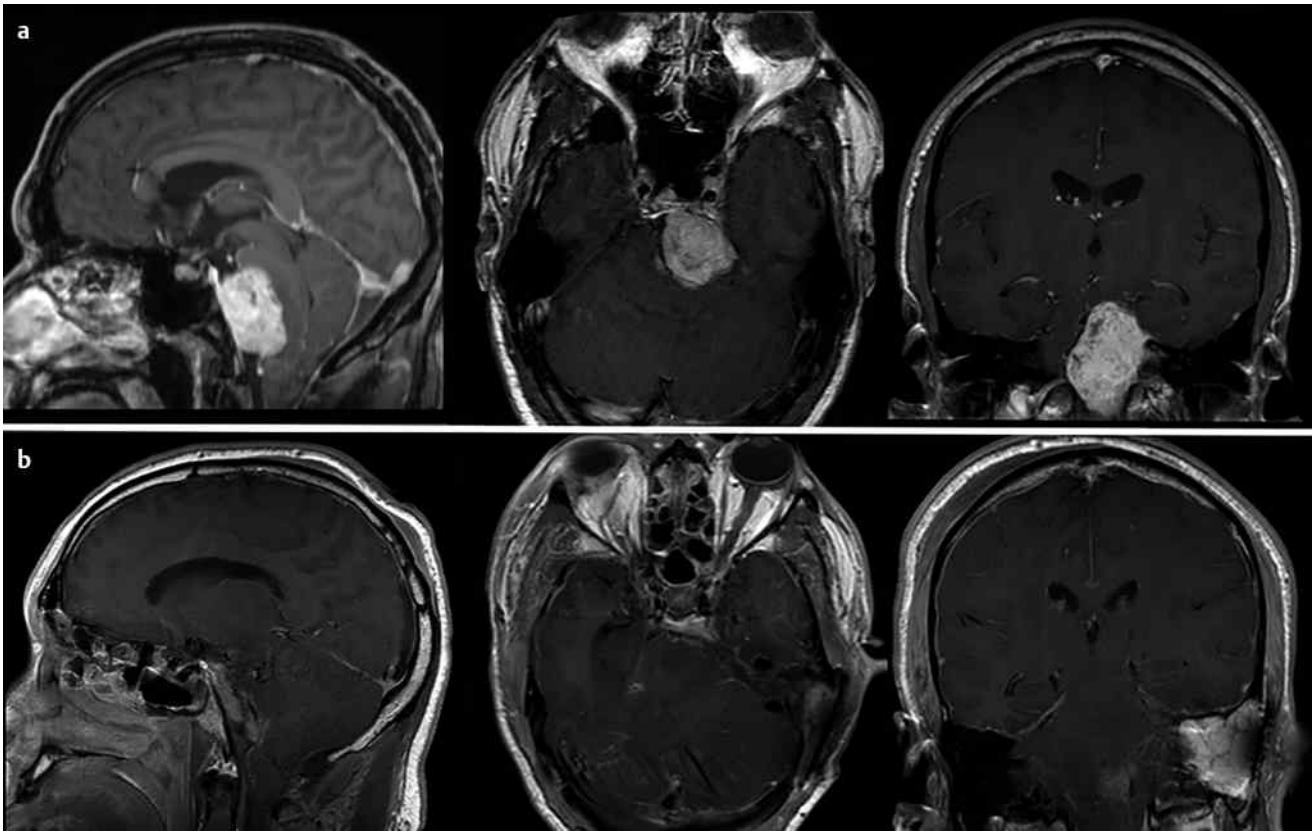


Fig. 13.11 Images of Case 2. (a) Preoperative images of a petroclival meningioma occupying zones 2 and 3, extending from the level just above the internal acoustic canal to the foramen magnum. This tumor was resected via a staged, multidirectional approach, combining a retrolabyrinthine posterior petrosectomy with a far-lateral transcondylar approach through the pre- and retrosigmoid corridors. **(b)** Postoperative images showed a near-total resection of the tumor.

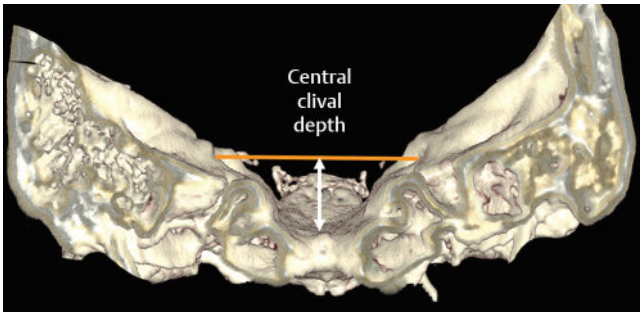


Fig. 13.12 View of the clivus from foramen magnum toward the dorsum sellae. The central clival depth is measured from the clivus to a line drawn between the internal acoustic canal on either side. For patients with deep drops, tumor attachments at the central clival depression are hidden from lateral and posterolateral approaches. These attachments may be best accessed via an endonasal endoscopic transclival approach.

magnum. Laterally, its reach is limited by the paraclival segments of the ICA, and the boundaries of the approach roughly correlates with lines in the parasagittal plane intersecting Dorello's canal (**Fig. 13.14**). Various modifiers increase the exposure from this "basic" form. An interdural pituitary transposition allows access to remove the dorsum sellae and posterior clinoid processes, thereby augmenting superior reach. Drilling the anterior petrous apex behind the carotid artery (with or without lateral mobilization of the artery itself) can increase lateral access to the superior petroclival region next to zone 1. Alternatively, a contralateral transmaxillary approach (via Caldwell–Luc or Denker's maxillotomy) can further facilitate the drilling of the petrous apex, offering a lateral reach not accessible through a purely endonasal approach. In zones 2 and 3, the "far-medial" extension of EEA allows the transclival approach to "flair out" laterally toward the jugular tubercle and occipital condyle (**Fig. 13.14**). Conservatively, using some of these modifiers, leaving the riskier maneuvers to endoscopic specialists, a "souped-up" endoscopic transclival approach can safely reach a petroclival meningioma in all three zones bilaterally, from the level of the trigeminal nerves, all the way to the level of the left and right hypoglossal nerves.

The endonasal transclival approach warrants particular consideration for reconstruction of the central skull base defect and separation from the nasopharynx. Full transclival defects pose a notably more complex reconstructive challenge than other EEA locations, in large part due to a significant dural defect as well as a high-flow leak with direct opening to the prepontine cistern. A modestly elevated incidence of postoperative CSF leak and even pontine herniation have been reported. Nuances vary by institutional preference, although the typical closure entails a multilayer reconstruction consisting of a dural substitute (or fascia lata), fat graft, and vascularized intranasal flap. The nasoseptal flap is particularly well-suited for local reconstruction given its large size, pliability, and ability to tolerate adjuvant radiation should it be required.

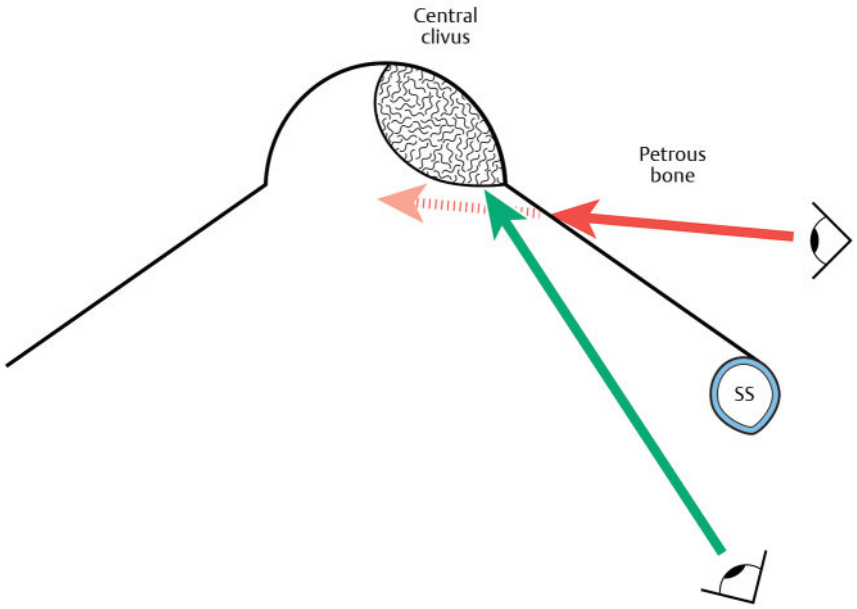


Fig. 13.13 Illustration of the tumor in the clival depression. Schematic drawing in axial orientation, showing that portions of the tumor attached to a deep clival depression is "blind" to approaches from the lateral trajectory such as via a presigmoid, transpetrosal. A retrosigmoid (posterolateral) trajectory may visualize this portion better, but the distance to target is very large. SS, sigmoid sinus.

Fig. 13.14 Ventral view of the extracranial surface of the clivus. The colored areas are reachable via the endoscopic endonasal approach (EEA). The *orange shaded area* denotes the "box" that can be access via the "standard" endoscopic endonasal transclival approach. The lateral borders roughly correlate with the parasagittal planes through Dorello's canal on either side. Superiorly, the "box" ends at the floor of the sella turcica and inferiorly at the foramen magnum. The *purple shaded area* indicates the region reachable with additional mobilization of the pituitary gland and drilling of the posterior clinoid process. The *pink shaded area* can be removed via the anterior superior petrosectomy through the EEA. The *green shaded area* denotes the regions accessible by further drilling toward the jugular tubercle and occipital condyles ("far-medial" extension of the EEA).

■ Combining Open and Endoscopic Approaches

It must be apparent at this point that the strengths and weaknesses of the "open" and endoscopic approaches complement each other perfectly. The "open" approaches from the lateral or posterolateral trajectory are excellent in reaching the petrous portions which are difficult to access through EEA's midline trajectory. On the other hand, the endoscopic approaches are perfectly suited to eliminate the meningiomas' clival dural blood supply early in surgery, and reaching the clival tumor components sometimes hidden from the "open" approaches. For right-handed endoscopic surgeons, who like to operate from the patient's right, petroclival meningiomas that are on the *left* side can even be resected *simultaneously* with combined "above-and-below" approaches. Whereas, for surgical teams that prefer staggering the timing of the two approaches, sometimes the time gap between stages can be surprisingly large.

Case Presentation 3

A 63-year-old man presented with several months of intermittent diplopia. He had similar symptoms 13 years prior, and was diagnosed with a left cavernous sinus meningioma. He was treated with radiosurgery, and his symptoms resolved. Lost to follow-up for many years, the MRI done for his recurrent symptoms showed a large petroclival meningioma, extending from the left posterior clinoid to the level of the midclivus (**Fig. 13.15**). A staged approach combining EEA transclival and anterior petrosectomy was designed, but after the EEA (**Video 13.2**), surgeons and patient all elected to postpone the anterior petrosectomy (stage 2). Two years hence, the tumor recurred again, and this patient finally underwent stage 2 for successful removal of his tumor.

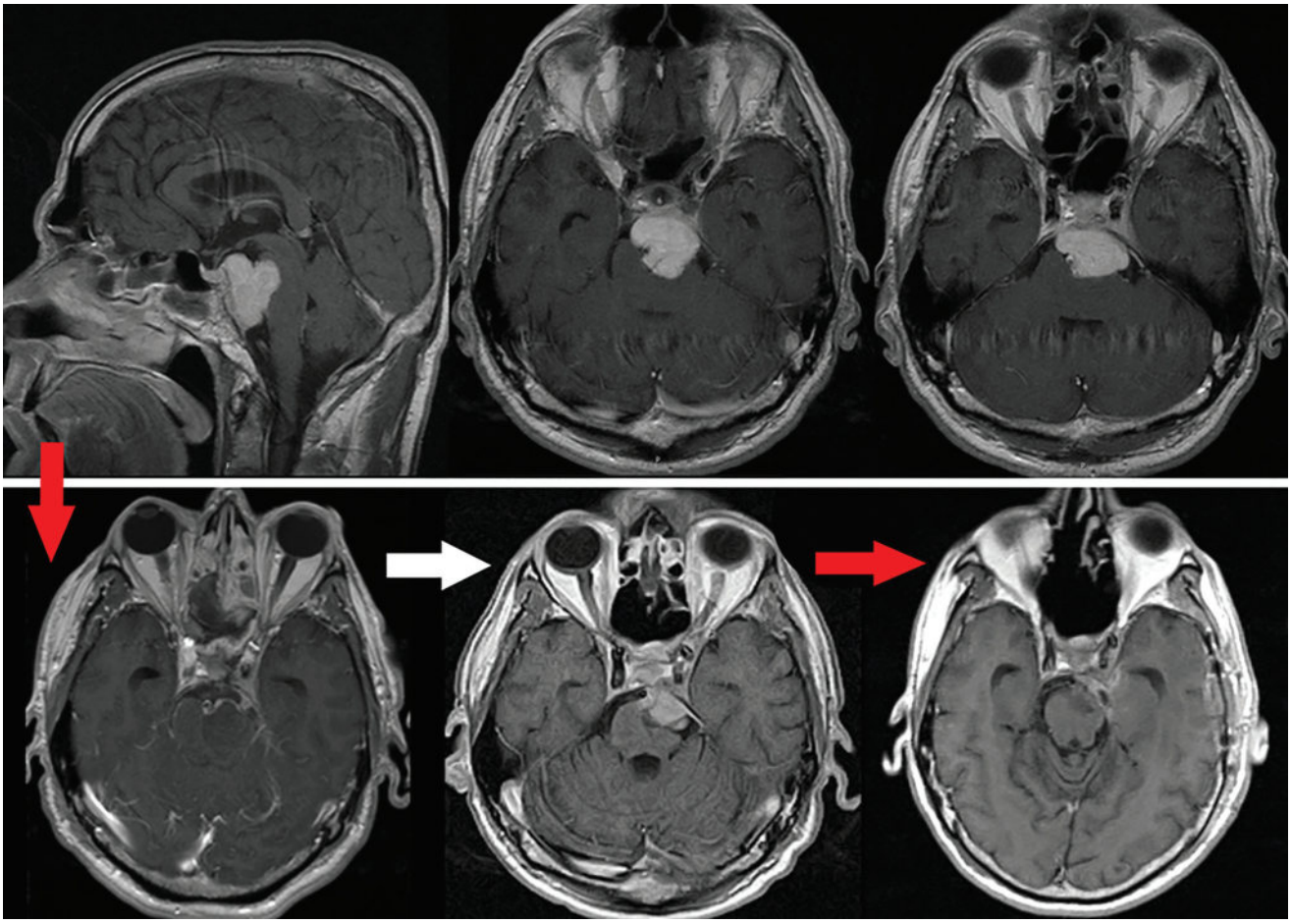


Fig. 13.15 Images of Case 3. Top row: images of the petroclival meningioma, which was treated with radiosurgery 13 years prior, when it was much smaller and contained in the left cavernous sinus. *Red arrows*, surgical intervention; *white arrow*, passage of time. The patient underwent EEA transclival resection (*down red arrow*), but after 2 years, the tumor once again regrew. An anterior petrosectomy (*horizontal red arrow*) was then performed for a partial resection.

■ Potential Complications and Associated Management

Common complications from endonasal endoscopic and microscopic transpetrosal approaches for petroclival meningiomas have been well-described, and they include CSF fistula, cranial neuropathies, brain injury related to venous infarction, and hydrocephalus. Brainstem injury can have a variety of causes and manifest in just as many forms. Direct injury from overzealous manipulation or dissection of an adherent tumor capsule from the brainstem can lead to problems ranging from motor deficits to difficulty with arousal. However, severe morbidities can also result from ischemic injury to the brainstem, which in turn can be traced to encasement of the basilar artery (BA) and its tributaries, large or small. This type of injury causes by far the most catastrophic neurological deficits, and its discussion is conspicuously underrepresented in the neurosurgical literature.

The majority of large petroclival meningiomas displace the main trunk of the basilar artery to the contralateral side (**Fig. 13.16**). However, it is far more common that these tumors, even small ones, would encase the ipsilateral posterior inferior cerebellar artery, anterior inferior cerebellar artery, or the superior cerebellar artery (**Fig. 13.1c**), depending on the location of the tumor in the clival zones. Disentangling these major branches from the tumor, though cumbersome, time consuming, and risky, is usually feasible with meticulous microsurgical technique (**Fig. 13.17**). On the other hand, in the rare occasion that the tumor truly encases the basilar artery in the 360-degree manner (**Fig. 13.16f**), its perforating branches that supply the brainstem must travel *through* tumor to reach its target, and disentangling these delicate vessels from the tumor may be completely futile and ultimately dangerous to the patient (**Fig. 13.17**).

Encountering this situation poses the modern neurosurgeon one of the greatest challenges in intraoperative decision making. Leaving the tumor between the BA and the brainstem would avoid damage to the perforating branches, but would there be too much tumor left and insufficient brainstem decompression? Taking the tumor between the BA and the brainstem might ensure adequate tumor removal, but would that not injure all the perforating branches and lead to a brainstem stroke? A potential answer lies in the *proportion* of the tumor that is anterior to the BA versus posterior to it (i.e., between the BA and the brainstem). If the vast majority of the tumor is *anterior* to the BA, and can be removed, then there is the possibility that the brainstem would be sufficiently decompressed, despite leaving the portion behind the BA as an intended residual. In

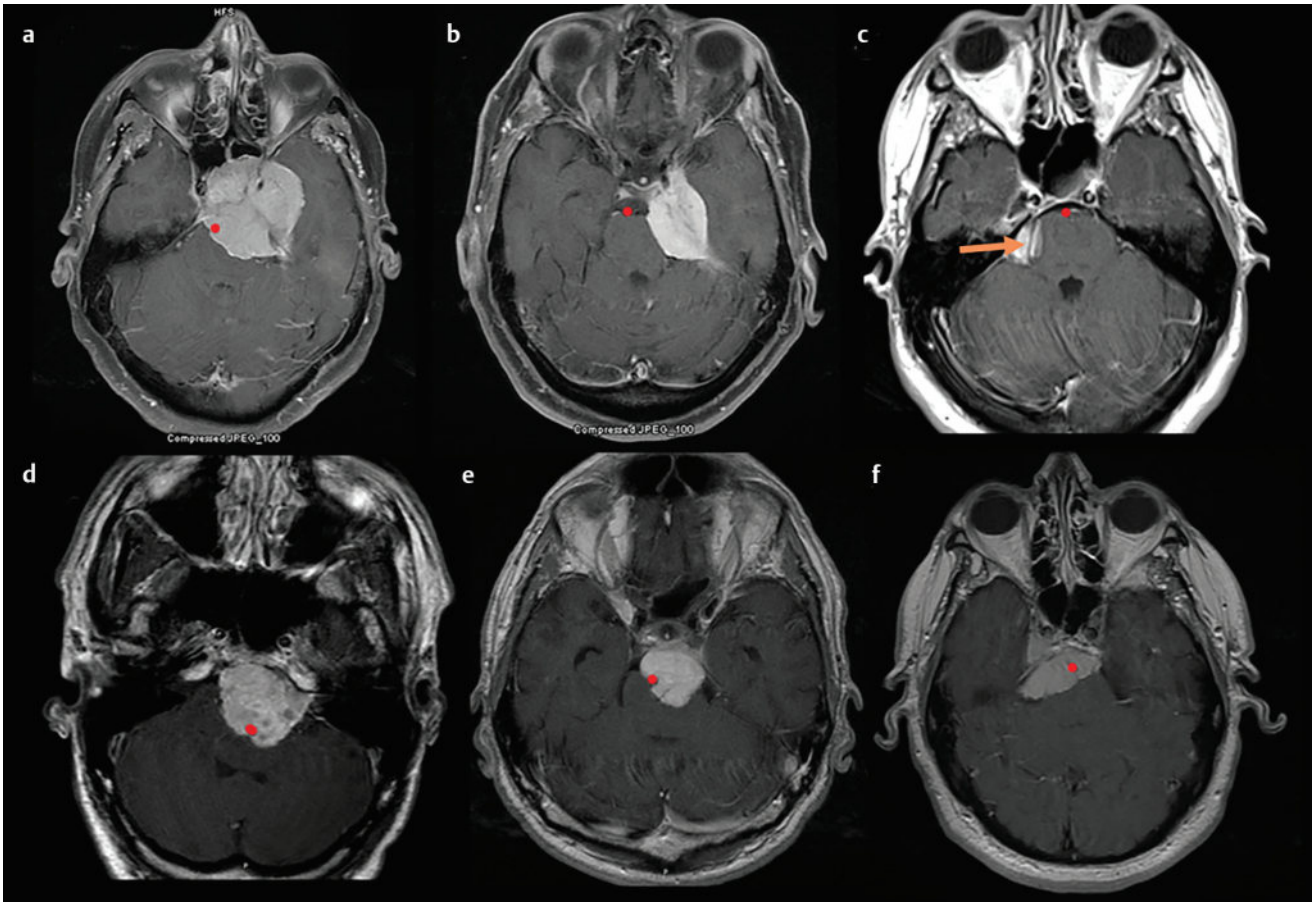


Fig. 13.16 Six petroclival meningiomas. Smaller tumors, such as **(b)** and **(c)**, may not involve in the basilar artery, but as shown in **(c)**, they can yet entangle major branches, such as the superior cerebellar artery (*orange arrow*). Large meningiomas, such as **(a)** and **(d-f)**, almost always displace the basilar artery to the contralateral side and the relationship between tumor and basilar perforating branches are variable. In **(f)**, the tumor surrounded the basilar artery completely, including multiple perforating branches.

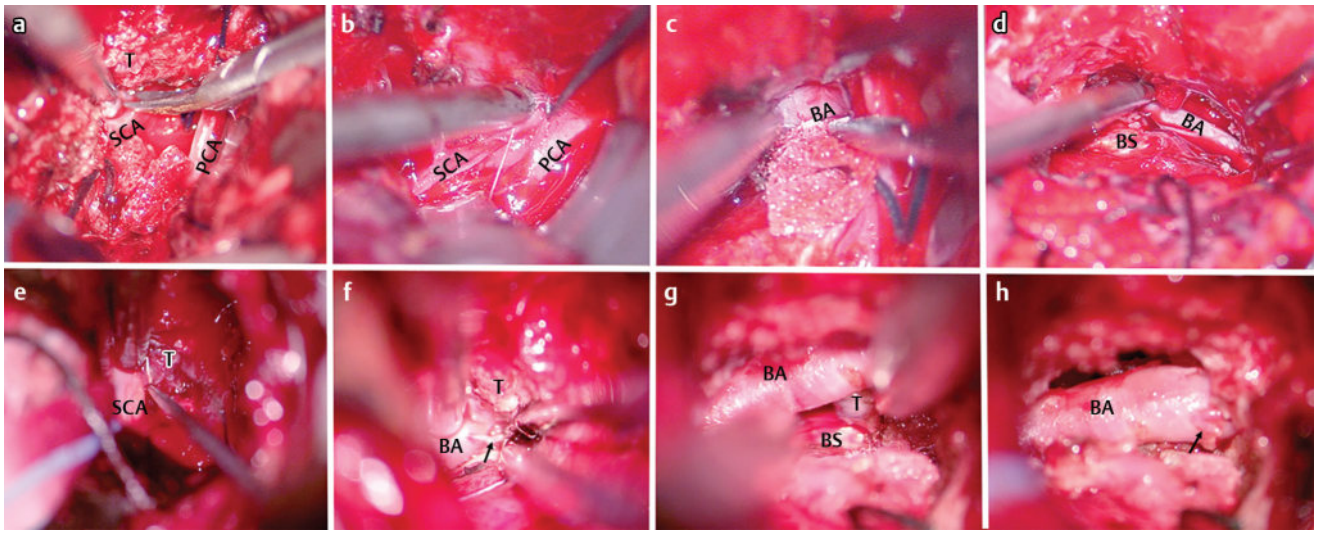


Fig. 13.17 Excavating the basilar artery (BA) in two cases. **(a–d)** Images from case 1, also shown in **Fig. 13.16a**. **(a)** The left superior cerebellar artery (SCA) was sharply dissected away from tumor, toward the basilar apex. **(b)** A sharp hook was used to excavate the SCA further toward the basilar apex. **(c)** The proximal basilar artery was found. **(d)** The BA was successfully disentangled from tumor. As shown, there was no tumor existing between the BA and the brainstem. **(e–h)** Images from case 4, also shown in **Fig. 13.16f**. **(e)** The right SCA was excavated from tumor. **(f)** Bleeding from a perforating artery was coagulated. (*arrow*, take-off of perforating artery from the BA). **(g)** Removing tumor in between the basilar artery and brainstem. **(h)** The basilar artery was disentangled from tumor which completely surrounded it. (*arrow*, the stumps of perforating arteries which were detached from tumor). BA, basilar artery; BS, brainstem; PCA, posterior cerebral artery; SCA, superior cerebellar artery; T, tumor.

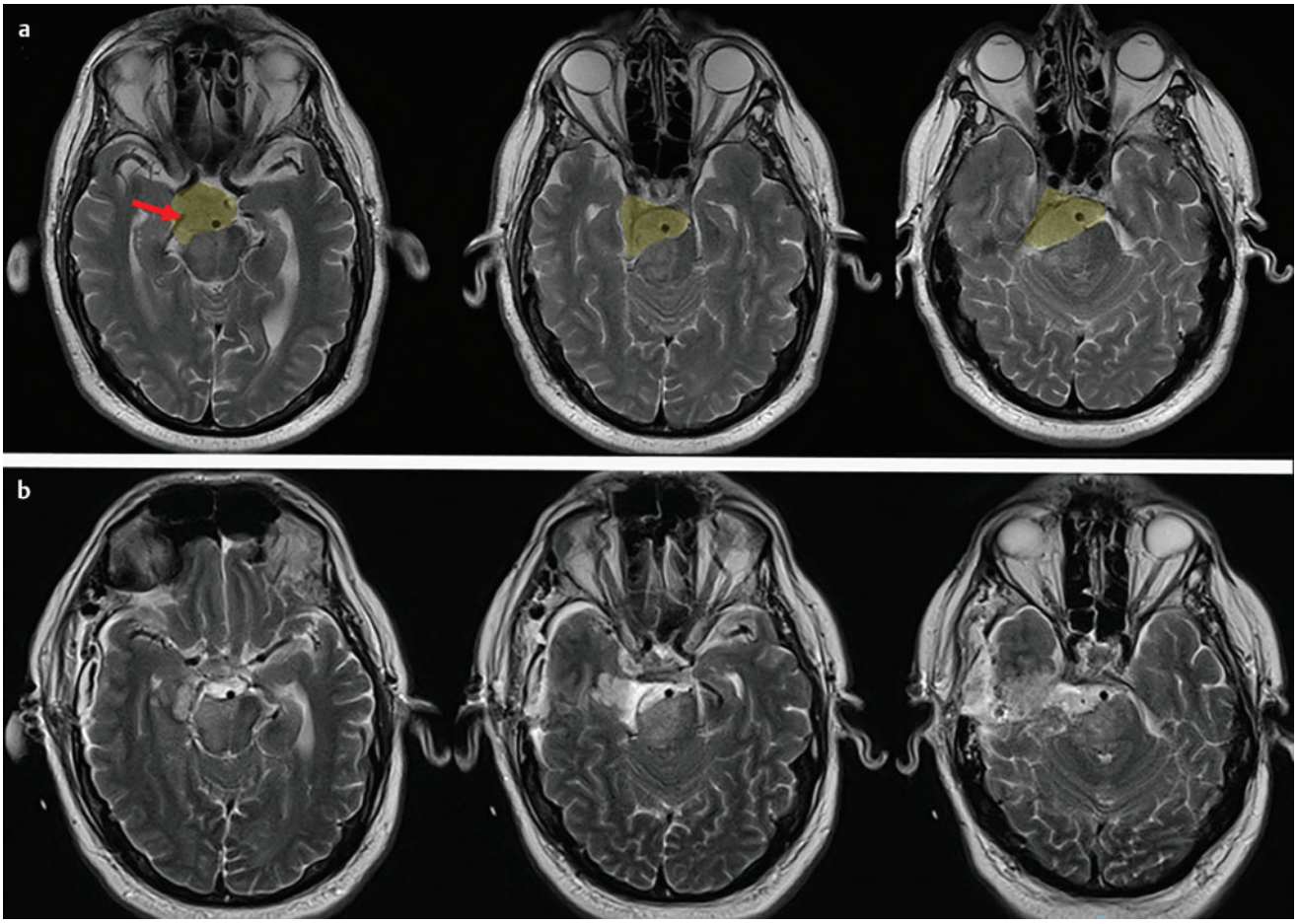


Fig. 13.18 Images of Case 4. (a) Preoperative and **(b)** postoperative T2 axial images of the patient's large petroclival tumor. The tumor is highlighted in faint yellow. *Red arrow* shows encasement of right posterior cerebral artery by the tumor. The basilar artery, seen as the *black dot* inside the tumor, was completely encased by the tumor throughout most of its course. Postoperative images showed an adequate resection, but the patient suffered a brainstem stroke from injury of the perforating branches of the basilar artery.

other words, it is a critical "judgement call" whether leaving tumor between the BA and brainstem as residual, thusly protecting the perforating branches, could still achieve the surgical goal, or whether it would lead to incomplete brainstem decompression. It is my personal creed to err on the conservative side.

Case Presentation 4

A 70-year-old woman presented with multiple falls in recent weeks. She had noted problems with her equilibrium for about 1 month. Her MRI showed a petroclival meningioma, occupying zones 1 and 2, but extending into zone 3 of the clivus, eccentric to the right (**Fig. 13.18**). Of note, both vertebral arteries end left of midline, and the BA also starts on the left and then becomes encased 360 degrees by the tumor for the rest of its course. A staged approach was designed, starting with a right anterior petrosectomy (stage 1). An EEA was intended for a second stage in order to find the vertebral arteries on the left, and to trace the BA from its healthy segment into the inferior and medial portion of the tumor. However, during stage 1, multiple perforating branches were found traversing the tumor, which was adherent to the brainstem. While a vast majority of the tumor targeted via the anterior petrosectomy was removed, the patient suffered a brainstem stroke after stage 1.

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14 Spheno-Caverno-Petroclival

Michaela Lee, Rami O. Almefty, and Peter Nakaji

Keywords: petroclival, cavernous sinus, cerebellopontine angle, surgical goal, quality of life

■ Case Presentation

A 72-year-old woman presented with the chief complaint of a hoarse voice. She had noticed changes in her voice and had been experiencing mild right frontal headache and blurry vision while reading for 2 months before seeing an otolaryngologist. On evaluation, her right vocal cord was paralyzed, and for this, she underwent computed tomography (CT) of the head and neck which showed an intracranial mass (**Fig. 14.1**). She was referred to our clinic for further evaluation. Neurological examination showed that all her cranial nerves were intact except for the vocal cord issue already mentioned. She had no balance problems or weakness. Magnetic resonance imaging (MRI) of the brain was then performed (**Fig. 14.2**).

Questions

1. What is the differential diagnosis based on all the imaging findings?
2. What are the management options? (Do you need any further imaging?)
3. If surgery is the selected therapy, what would be your goals for this 72-year-old patient?

■ Diagnosis and Assessment

MRI of the brain with and without contrast demonstrated a large, right-sided homogenously enhancing petroclival extra-axial mass expanding the prepontine cistern and extending into Meckel's cave, cavernous sinus, sella, anterior wall of internal auditory canal (IAC), and jugular foramen, producing mass effect on the brainstem. The MRIs revealed no obstructive hydrocephalus and minimal vasogenic edema. The cavernous carotids were relatively equal in size bilaterally. The tumor had grown around the basilar artery, almost circumferentially, and thus did not displace the artery significantly. The vicinity of the tumor to the lower cranial nerves appeared to be the cause of her symptoms. A second, smaller mass that was located superiorly was consistent with a planum sphenoidale meningioma. Although the second mass was potentially an extension of the petroclival tumor, given its shape and resemblance to a "classic" planum sphenoidale meningioma, we interpreted the imaging findings as two separate masses. The patient also had other very small meningiomas over the left frontal, occipital, and right temporal convexities, which were of little clinical significance on their own; thus, we believed that these small meningiomas could be followed with surveillance imaging. On the limited CT slices that were available, reactive changes to the primary tumor could be seen in the petrous bone and clivus.

Meningioma was the most likely diagnosis, but one could also consider a schwannoma, a chondrosarcoma, or metastasis. Meningiomas are generally slow growing and symptoms are usually insidious in onset. On MRI, they can have a classic dural tail and sometimes, as in this case, a cleft of cerebrospinal fluid (CSF) around the tumor can be seen, which denotes separation of tumor from the surrounding parenchyma. Although it was not needed for this patient, preoperative CT of the temporal bone and MR venography can be useful in surgical planning to understand the location of the tumor in relation to certain structures such as the IAC, jugular bulb, transverse and sigmoid sinuses, and large draining veins.

We determined that the petroclival mass was most likely the cause of the patient's symptoms and that surgical resection would provide both a diagnosis and immediate decompression of the nerves and brainstem. The planum sphenoidale mass can be addressed, if necessary, during an operation in the future, as both lesions cannot be safely removed in one simultaneous operation.

The age and medical condition of the patient should always be carefully considered in formulating a surgical plan. In this case, the patient's symptoms and the need for brainstem decompression were weighed carefully against the potential risks of all the surgical options and within the context of her overall condition and life expectancy. Our patient's main symptom was voice hoarseness. She was otherwise a highly functional individual, working full time and leading an active

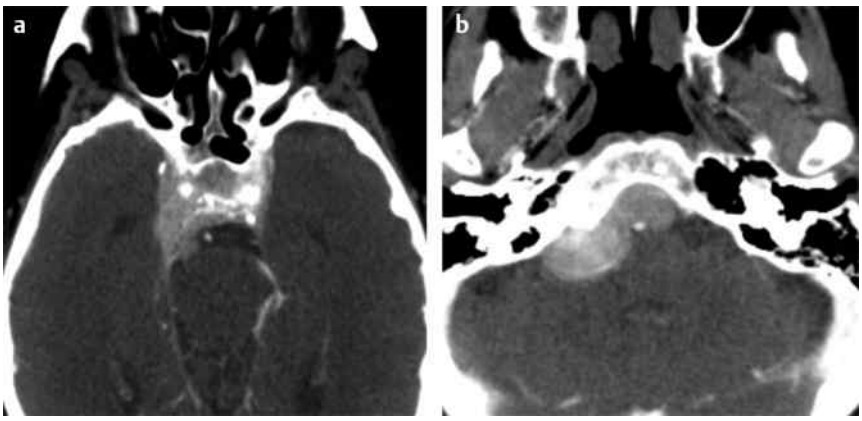


Fig. 14.1 (a, b) Axial computed tomograms with contrast show a hyperdense mass in the petroclival region extending up to the cavernous sinus and Meckel's cave. Hyperostotic changes of the clivus are also notable on the right side. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

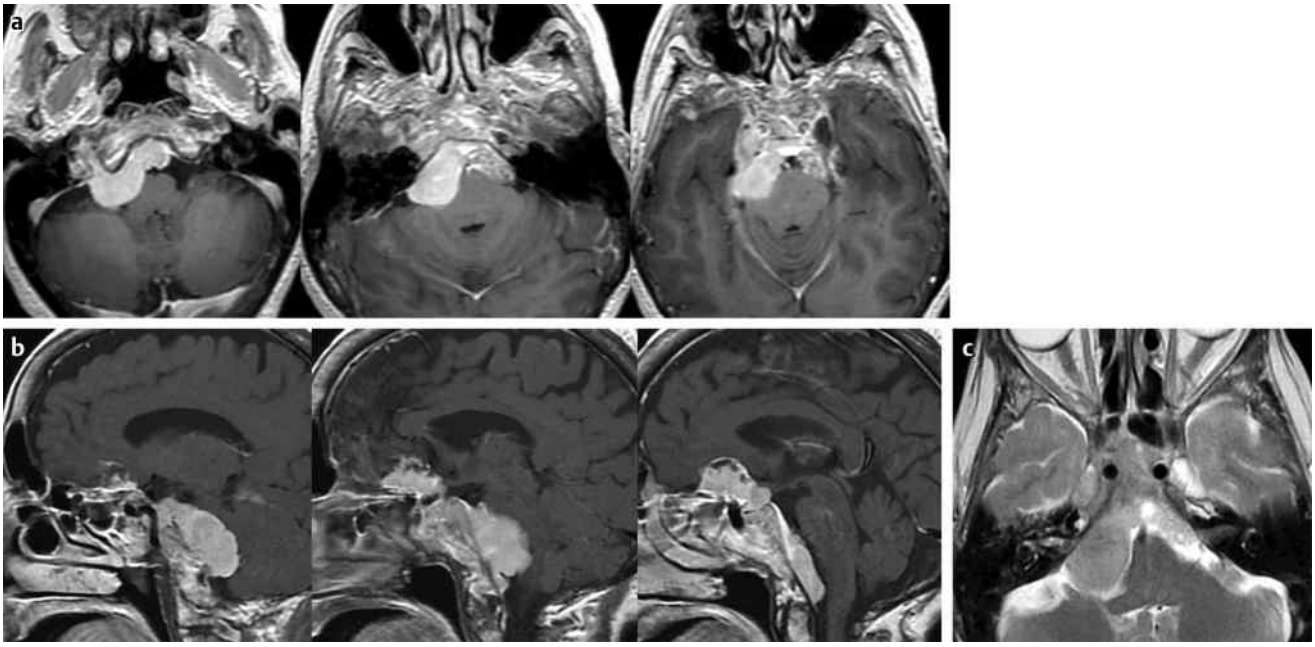


Fig. 14.2 Serial **(a)** axial and **(b)** sagittal T1-weighted magnetic resonance imaging (MRI) with contrast and **(c)** axial T2-weighted MRI demonstrate an avidly enhancing extra-axial petroclival mass with brainstem compression. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

life. The imaging studies showed the petroclival mass that was quite irregular and involved multiple vessels and cranial nerves. Complete resection would be very difficult, without a high risk of leaving the patient with major neurological deficits. Therefore, we set the surgical goal for brainstem decompression, with a maximally partial resection without iatrogenic neurological injury. By limiting the goal, we hoped to maintain her quality of life, even at the expense of leaving some tumor behind. As these meningiomas grow but slowly, any residual tumor can be observed with surveillance imaging or treated with adjuvant radiotherapy. The ethos of this strategy does not provide the surgeon a "license" to be lazy or careless; the tumor must be resected as thoroughly as possible, with meticulous dissection and safeguard of all critical structures. It simply recognizes that maintenance of the quality of life trumps a complete resection with a "clean" postoperative image.

■ Anatomical and Therapeutic Considerations

Surgical resection of petroclival meningiomas can be challenging and technically demanding. These tumors usually arise near the spheno-occipital synchondrosis, anterior to the IAC. They can grow quite large, extending to the tentorial incisura, middle fossa, cavernous sinus, and posterior fossa. Often by the time a patient experiences symptoms, the tumor has started to displace the brainstem and cranial nerves and it often encases critical neurovascular structures. Because of a combination of these factors, gross total resection can be particularly difficult to achieve without significant morbidity for the patient. In addition to the tumor's encasement of critical neurovascular structures, the absence of an arachnoid plane between the tumor and brainstem with pial infiltration or hyperintensity in the brainstem on T2 fluid-attenuated inversion recovery MRI can also make gross total resection nearly impossible without unacceptable morbidity. Extensive knowledge of the complex anatomy of the involved region is therefore imperative for the surgeon who tackles these tumors.

Patients with petroclival meningiomas usually present with worsening headaches, with or without obstructive hydrocephalus, cranial nerve or cerebellar dysfunction, and symptoms related to brainstem compression. Our patient presented with lower cranial nerve dysfunction. Brainstem compression also frequently causes significant neurological deficits; therefore, decompression of the brainstem is usually one of the primary goals of surgery.

In determining the surgical approach, the surgeon should consider several patient and tumor characteristics: preoperative Karnofsky Performance Scale score, invasion into the cavernous sinus, cranial nerve involvement, brainstem compression and edema, encasement of critical vessels, and vascularity of tumor and feeders from the vertebrobasilar system. For some patients, preoperative embolization to reduce intraoperative blood loss may be appropriate, as petroclival tumors are often supplied by branches of the middle meningeal artery, the tentorial artery, or the ascending pharyngeal artery. Moreover, depending on the complexity of the tumor and the combination of approaches (i.e., anterolateral and posterolateral), surgeries can be staged.

Options for Approach

Historically, petroclival meningiomas have been resected using some form of transpetrosal approach or a variant thereof. For instance, an anterior petrosal approach (e.g., the Kawase approach) allows access to lesions in the upper petroclival region from the oculomotor nerve (cranial nerve III) to the IAC

by providing a window to the anteromedial cerebellopontine angle, petrous apex, Meckel's cave, and ventrolateral brainstem. For this patient, an anterior petrosectomy would not provide adequate exposure to the posterior petrous region and inferior half of the clivus.

Posterior petrosal approaches, such as the retrolabyrinthine presigmoid, translabyrinthine, and transcochlear approaches, generally aim for the region posterior and lateral to the IAC. A retrolabyrinthine approach has the advantage of preserving hearing, but it provides limited access to the petroclival region by itself. Like many of the approaches mentioned above, it is more useful in combination with a middle fossa craniotomy. A translabyrinthine approach gives access to the space between the tentorium and jugular foramen and, although it might be an option for this patient, it would damage her hearing. It is also difficult to reach the lower cranial nerves IX–XI (i.e., the glossopharyngeal, vagus, and spinal accessory nerves) with this approach. The most aggressive approach on this spectrum is the transcochlear approach. This provides the widest exposure, from the pons to the medulla, and allows one to reach lesions ventral to the brainstem. However, it requires facial nerve transposition, which would leave the patient with postoperative facial weakness.

In summary, aggressive approaches such as the translabyrinthine or transcochlear approach would provide sufficient exposure to remove our patient's tumor but at the expense of hearing loss and facial paralysis. Total petrosectomies have thus fallen out of favor with many surgeons because of the significant morbidity associated with these approaches and, even with maximal exposure, complete resection cannot be guaranteed. There will always be some part of the dural attachment at the skull base that cannot be removed, and tumor cells are therefore left behind even with a large exposure. Because of these potential complications and outcomes, in many cases, the surgical benefits of complete resection do not outweigh the significant morbidity that comes with it.

Approach of Choice

Given the advanced age of our patient and the size of her tumor, she would likely not have tolerated a long operation with maximal exposure without experiencing some postoperative neurological deficits. Because our surgical goal was maximally safe resection of the tumor, explicitly for brainstem decompression, and not for total resection, we opted for a staged approach, starting with a right retrosigmoid craniotomy, leaving the left side of the petroclival tumor, and the planum sphenoidale tumor, for a later stage if needed.

The retrosigmoid approach is one of the workhorses of neurosurgery, as it provides exposure to the posterior surface of the petrous bone, ventrolateral brainstem, and foramen magnum. It is versatile and can be modified with as much or as little bony removal as is needed to obtain adequate exposure. For example, a far-lateral approach can be combined with a retrosigmoid approach for a more inferior-to-superior anterolateral exposure.

This approach would provide adequate exposure in our patient to achieve the goal of brainstem decompression, without compromising her hearing and facial nerve function. The T2-weighted images showed an arachnoid plane between the tumor and the basilar artery, which would facilitate tumor removal. Although it may be difficult to reach the anterior and contralateral part of the brainstem as one is limited by the facial and vestibulocochlear nerves (cranial nerves VII and VIII), removal of the tumor in these areas may not be necessary to accomplish the goal of brainstem decompression.

■ Description of the Technique

Our patient was positioned supine with a right shoulder bump and her head turned to the left, placed in a Mayfield head holder, and secured to the operating table. Neuromonitoring of cranial nerves VII–XII (facial, vestibulocochlear, glossopharyngeal, vagus, spinal accessory, and hypoglossal nerves) was set up and baseline values were obtained (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: inferior, posterolateral

Craniotomy: retrosigmoid

Modifier: none

Neuronavigation was registered and accuracy was confirmed. It was then used to delineate the transverse–sigmoid junction (TSJ) and a slightly curvilinear retroauricular incision was marked out. The patient was then prepped and draped in a sterile fashion and the incision was made sharply down to bone. After the location of the TSJ was reconfirmed with neuronavigation, a burr hole was placed just inferior and posterior to the junction. Then a craniotome with a footplate attachment was used to turn the craniotomy flap, staying below the transverse sinus and behind the sigmoid sinus. All exposed mastoid air cells were waxed. The dura was opened and tacked up, and the operating microscope was brought into the operative field (see **Operative Setup**).

The cerebellopontine angle cistern was opened sharply to allow egress of CSF and to relax the cerebellum. Microscopic techniques and sharp dissection were used to carefully dissect

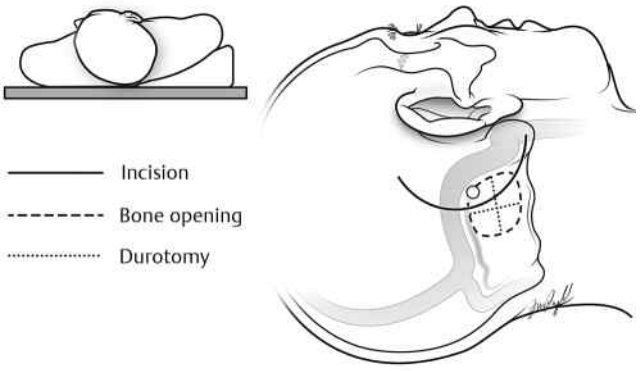
Operative Setup

Position: supine, head turned to left

Incision: retroauricular

Bone Opening: retrosigmoid

Durotomy: cruciate



the cerebellum away from the tumor (**Fig. 14.3a**). We then proceeded to devascularize the tumor by disconnecting it from the posterior petrous surface using bipolar coagulation and sharp dissection. We identified cranial nerves VII–XI and, using a nerve stimulator, stimulated the lateral surface of the tumor (**Fig. 14.3b**). After confirming that the lateral surface was devoid of any of nerves, we began the initial debulking of the tumor with an ultrasonic aspirator (**Fig. 14.3c–e**). A small specimen was sent for frozen pathological examination, which confirmed a diagnosis of meningioma. Once the tumor was significantly debulked, we began rolling the tumor and dissecting it away from the cerebellum and cranial nerves VII, VIII, IX, and X.

With continued resection, we found a favorable plane between the tumor and the brainstem and peeled it away without compromising the brainstem or its vasculature. As we proceeded more inferiorly, we dissected the tumor away from cranial nerves IX, X, and XI where they entered the jugular foramen, as well as away from cranial nerve VI as it proceeded toward the Dorello canal. We removed the tumor from the clivus as much as possible but, as expected, could not reach the contralateral side without placing traction on the brainstem.

We then turned our attention to the lower medullary component, where we removed tumor from cranial nerve XII (**Fig. 14.3f**). At this point, we had removed as much tumor as we safely could using this approach (**Fig. 14.3g**). After hemostasis was obtained, the surgical field was irrigated copiously with antibiotic-impregnated saline. The dura was closed with a bovine pericardium patch to achieve a watertight closure and a dural sealant was applied. We again waxed any exposed air cells and replaced the bone flap with a small piece of mesh using titanium plates and screws.

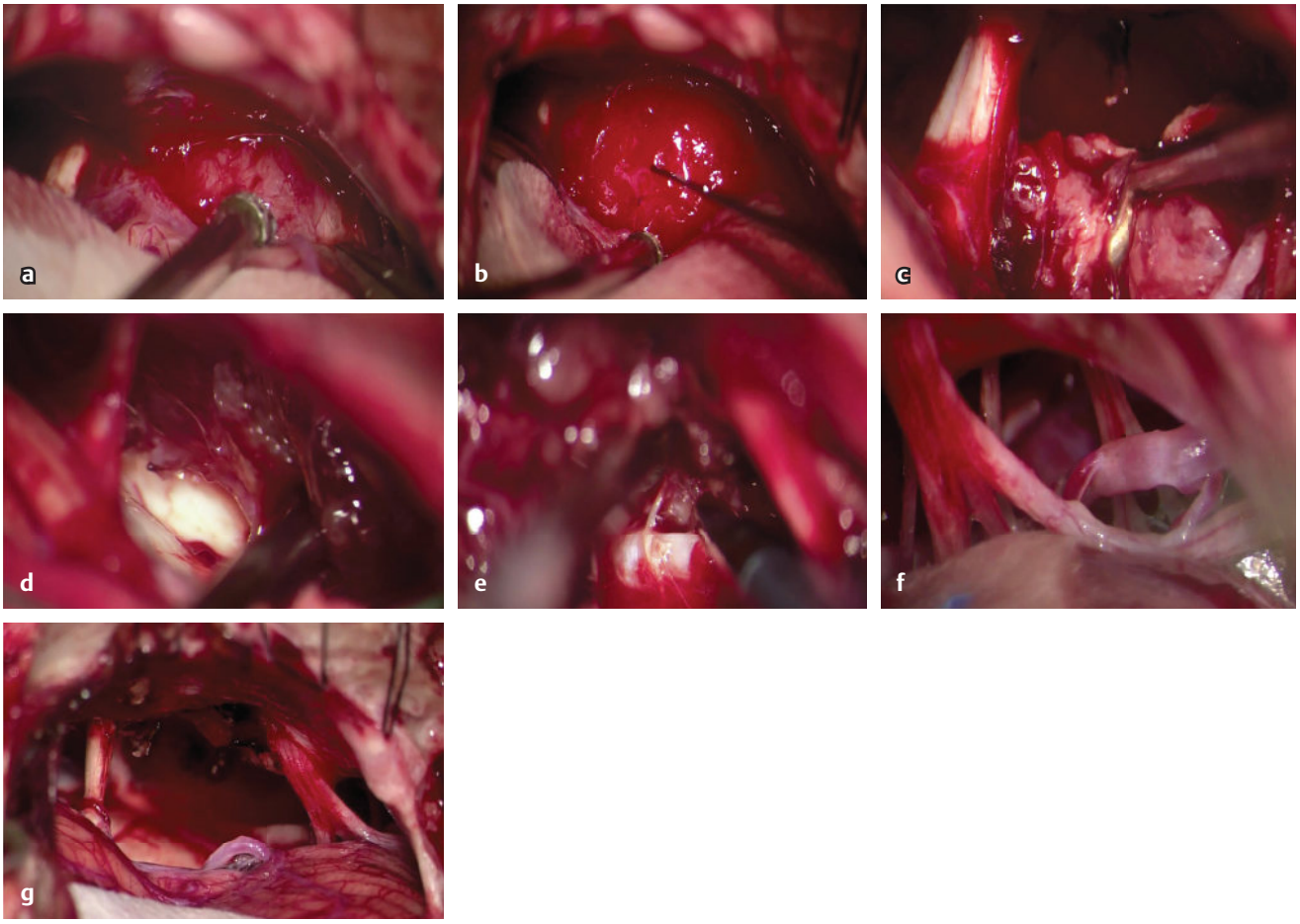


Fig. 14.3 Intraoperative views. **(a)** Tumor (*middle*), cerebellum (*bottom*), and cranial nerve VII–VIII complex (*left*). **(b)** Nerve stimulator on tumor capsule. **(c)** Debulking of tumor with the cranial nerve VII–VIII complex to the left. **(d)** Intraoperative view of brainstem during tumor debulking and dissection. **(e)** Dissection of tumor off the basilar artery. **(f)** Cranial nerves IX–XII and the posterior inferior cerebellar artery (*center*). **(g)** Resection cavity with cranial nerve VII–VIII complex to the left, cranial nerves IX–XI to the right, anterior inferior cerebellar artery in bottom center, and basilar artery in deep center of picture. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

Surgical Pearls

1. With the advent of radiosurgery as a viable adjuvant treatment for small residual tumors, the treatment paradigm has shifted toward maximal partial resection without iatrogenic injuries and away from maximal resection at any cost. Limiting the surgical goal and resection to maintain the quality of life is particularly reasonable in an older patient, who may not be able to tolerate a long and aggressive surgery, and whose natural lifespan may limit any damage a residual tumor can cause.
2. Although it is important to know the anatomical landmarks, such as the asterion, as demarcation of the junction of the transverse and sigmoid sinuses, neuronavigation can be helpful in planning the craniotomy because of variability in individual patient anatomy.
3. For a retrosigmoid approach, flexing the neck of the patient and then slightly angling the vertex down toward the floor will maximize the surgeon's view and comfort during the resection of the superior part of the tumor. It is often helpful to have the patient mimic the positioning preoperatively to see if the neck position is tolerable.
4. Devascularization of the tumor by separating it from the skull base is a good first step.
5. Tumor debulking to reduce the mass should be undertaken to facilitate separation of the tumor–brainstem and tumor–cranial nerve interfaces.
6. The surgeon should take intermittent mindful pauses during surgery to consider whether the current strategy or goals should be redefined.

■ Aftercare

Given the size and location of the tumor and its involvement with the cranial nerves, our patient did well after surgery. Postoperative MRI demonstrated sufficient decompression of the brainstem and a small residual tumor in the contralateral prepontine cistern, Meckel's cave, and cavernous sinus (**Fig. 14.4**). Although the total percentage of the tumor removed was modest, the amount that was excised corresponded to what we had intended to remove at the beginning of the surgery.

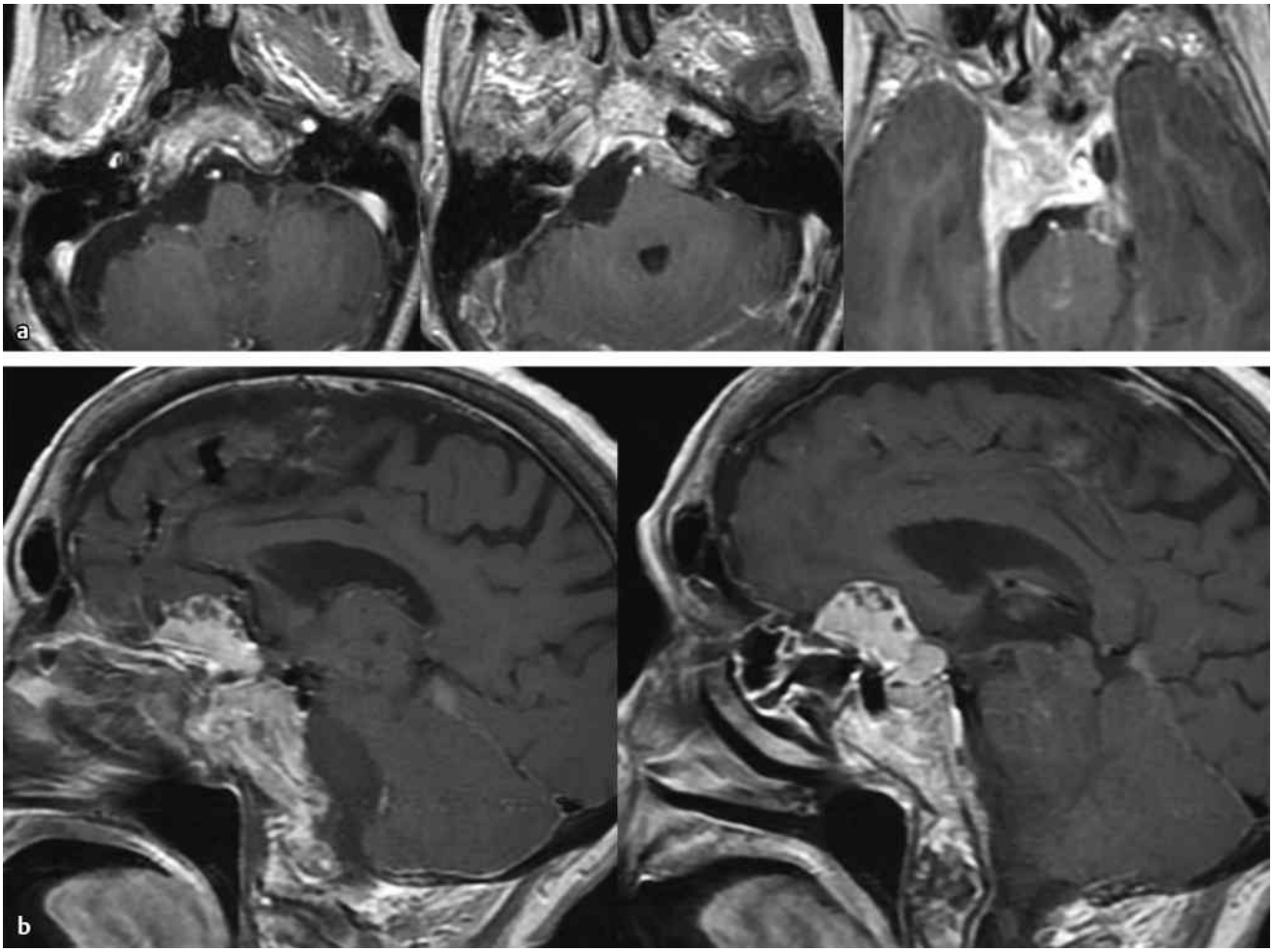


Fig. 14.4 Postoperative serial **(a)** axial and **(b)** sagittal T1-weighted magnetic resonance images with contrast demonstrate good decompression of the brainstem and a small but expected residual in the contralateral cistern and cavernous sinus. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

The patient had a right cranial nerve VI palsy that was not unexpected, as the tumor was adherent to that nerve and it was manipulated off the surface of the tumor during surgery. We expected this deficit to resolve over time and by postoperative day 4, the palsy was already improving. A follow-up laryngoscopy once again showed the right vocal cord paralysis and an incomplete compensatory closing by the left vocal cord. The rest of the cranial nerves remained intact. The patient was discharged to acute rehabilitation on postoperative day 5 and returned home shortly afterward. Her voice also improved after a vocal cord injection by the otolaryngology service.

The histopathological diagnosis was transitional meningioma World Health Organization grade I with an MIB-1 index of 2%. Given the low grade of the tumor and its stability on 3-month follow-up imaging, we elected to follow the patient with serial imaging and delay the second-stage operation. The primary surgical goal had been achieved, and the patient was satisfied with the result.

■ Possible Complications and Associated Management

Petroclival meningiomas are challenging tumors because of their involvement with critical neurovascular structures. No two tumors are exactly the same. Some are soft and separable, whereas others are quite vascular, unyielding, or tightly enmeshed with the nerves and vessels. When the tumor is unfavorable, cranial nerves can be stretched or injured, causing temporary or permanent dysfunction. Although meticulous techniques are always critical, it is often better to leave some tumor behind rather than causing a permanent iatrogenic injury. Transient diplopia due to manipulation of cranial nerve IV or VI is common, but fortunately it is also usually temporary, and a short course of corticosteroids can also help reduce any irritation. If there is any concern for cranial nerve IX, X, or XII dysfunction, a swallow evaluation is needed after extubation prior to starting the patient on any oral intake. Vocal cord dysfunction can also be temporarily managed with a vocal cord injection.

As with any neurosurgical procedure, one must respect the vessels during petroclival tumor resection. Depending on the tumor, petroclival meningiomas can be extremely adherent to large vessels such as the basilar artery and its perforators. Injury to any traversing vessel can cause an ischemic stroke. Brainstem injury can also occur if the pial surface is violated during surgery; therefore, any tumor that is adhered to the brainstem and its pial vessels should be left alone.

For posterior fossa craniotomies, the risks of postoperative hemorrhage and CSF leak are more concerning than they are for supratentorial craniotomies. The posterior fossa is also much smaller and less tolerant of any space-occupying hematoma. Meticulous hemostasis should be achieved prior to closing, and blood pressure goals should be set and maintained within a normal range postoperatively. With a posterior fossa craniotomy, closure of the dura must be watertight to reduce the risk of CSF leak. Nevertheless, if a leak occurs despite one's best efforts, treatment options include elevating the head of the patient's bed, placing a lumbar drain, reexploring the wound for primary repair, and placing a shunt.

Perspective

Pankaj K. Agarwalla, R. Tushar Jha, Siviero Agazzi, and Harry R. van Loveren

■ Introduction

In the preceding section, the authors discussed a highly complex tumor that reached from the planum sphenoidale all the way to the foramen magnum. And yet, they were able to formulate a clear plan, perform the operation to achieve the surgical goals, and discharge the satisfied patient to rehabilitation 5 days after surgery. The key to this excellent outcome is a set of surgical goals which were set judiciously, that in turn led to an approach which was rational. While the authors described their management as well as some of the philosophical underpinnings of their decision making, we will expand on some of these philosophical concepts, since they are often as critical as the technical details of surgery.

Whereas it is impossible to prescribe one approach for each tumor, the choice should be guided first and foremost by the surgical goal formulated for each individual patient. This is not to say that the plethora of nuances, anatomical and clinical, is unimportant, but at the very start of the decision-making process, these are distractions. With the guiding principle that the surgical goal must be set first, one cuts through a lot of "noise" which unnecessarily complicates the process at this stage.

■ Setting the Surgical Goals

For setting the goals, we recommend discussing the goals of surgery both with the patient and with your colleagues, such as in a tumor board setting. A discussion of the goals of surgery, particularly with the patients, will emphasize the risks they are willing to undergo and also give you a chance to understand what the patient hopes to gain from any surgical resection.

Case Presentation 1

A 42-year-old woman presented to our clinic with an 18-month history of headaches, blurry vision, and a complex history of a cavernous sinus meningioma. On initial presentation about a year prior, an MRI revealed a holocavernous meningioma. While under the care of a separate neurosurgical team, she underwent an endoscopic transsphenoidal biopsy that demonstrated a grade III meningioma and was subsequently treated with fractionated radiation therapy. A month prior to her current presentation, she developed worsening headaches and progressive right ophthalmoplegia. A follow-up MRI was obtained and demonstrated marked enlargement of her tumor **(Fig. 14.5)**. Her neurological examination was notable for right cavernous sinus syndrome including diplopia, complete right cranial nerves IV and VI palsies, a partial cranial nerve III palsy with ptosis, and decreased sensation to light touch in the V1–V3 dermatomes.

The patient and her family understood the aggressive nature of this tumor and sought appropriate treatment to prolong survival. Emphatically, she was willing to risk permanent deficits for this aim, and even specifically stated that if we needed to "take her eye," that would be acceptable. She understood clearly that any attempt at surgery for a meaningful resection would involve sacrifice of the right internal carotid artery and no useful function of her right eye. Having consented for the surgery, the patient underwent successful balloon test occlusion and coil embolization of the right internal carotid artery **(Fig. 14.5c–e)**. In a separate setting, she underwent a right frontotemporal orbitozygomatic (FTOZ) craniotomy and anterior clinoidectomy for complete resection of the malignant holocavernous meningioma.

This case illustrates how "patient factors" can dictate the process of setting the surgical goal, which in turn determines the unique surgical approach. Certainly, resection of holocavernous meningiomas is not routinely performed in our practice, but whereas "patient factors" limited the goals for Dr. Nakaji's patient, they expanded them in the example above. In both situations, the goals ultimately determined the right surgical approach for each patient.

■ Strategies to Achieve the Goals

Once the goals of surgery are set, then you must choose the most straightforward path for achieving them. It is important to emphasize that, whereas "long-term tumor-free survival" is *sometimes* the goal, getting a "clean" postoperative scan, one that you can show as a trophy to your colleagues, is *never* the goal. Here, we are reminded that more complex approaches that might achieve a "clean" scan invite "operative-time-dependent" risks, including deep venous thrombosis, urinary infections, pulmonary embolism, infection, and wound healing problems to name only a few. Like Dr. Nakaji's case, we advocate for more straightforward approaches, such as the well-done retrosigmoid craniotomy for petroclival meningiomas, if this can achieve the goal of surgery, which in his case was brainstem decompression first and foremost.

This is not to say that complex approaches are useless—quite the opposite! Complex skull base approaches can provide critical corridors for better exposure, tumor resection, and protection of critical structures when the surgical goal requires all three. However, as each element of a complex plan is added piece-by-piece to a growing strategy, you must be clear how each element improves the exposure, tumor resection, and control of the neurological surrounding.

Once a plan is out of the design phase, the surgical goals serve two purposes during the execution phase. The first is to tell us when to stop the operation. It is absolutely critical that during surgery, you are reminded of the goals of surgery and stick to the general plan to avoid becoming "greedy" and taking unnecessary risks, particularly when this goes against patient's expectation of surgical risk-taking. Never forget the option to stop and allow yourself the chance to come back and fight another day.

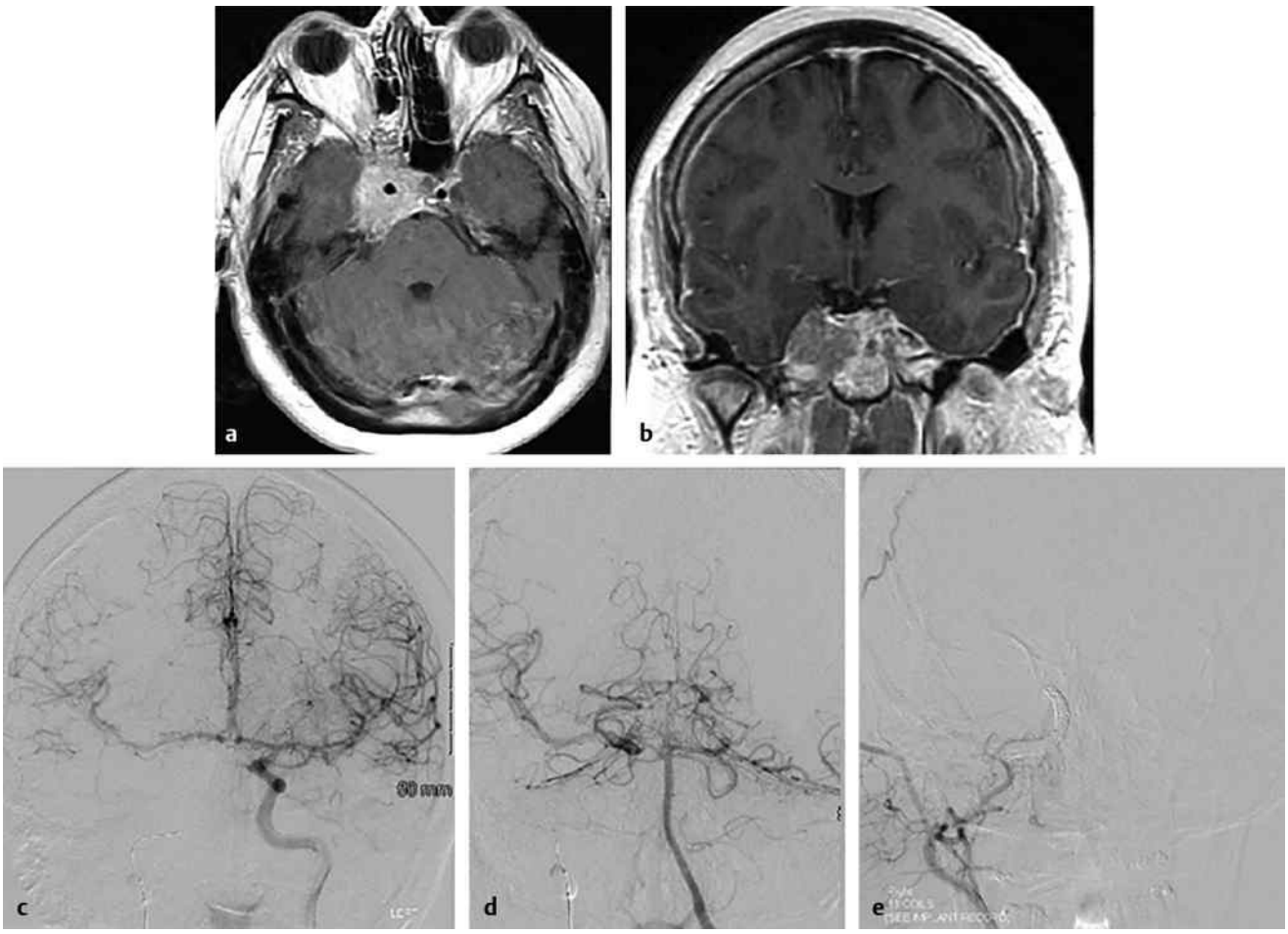


Fig. 14.5 Images of Case 1. (a) Axial and **(b)** coronal T1 MRI after gadolinium administration demonstrating a 2.5 × 3.3 × 3.1 cm right holocavernous meningioma. The tumor was confined within the lateral wall of the cavernous sinus, encasing the right internal carotid artery and cranial nerves. **(c, d)** Cerebral angiogram demonstrated successful balloon test occlusion (BTO) of the right internal carotid artery (ICA). Anterior–posterior projections of cerebral angiogram during BTO of the right ICA and contrast injection of the left ICA and left vertebral artery (VA), respectively, demonstrated filling of the right anterior cerebral artery (ACA) and middle cerebral artery (MCA) via blood flow from the anterior communicating artery (Acom) and posterior communicating artery (Pcom). **(e)** Anterior–posterior projection following coil embolization of right ICA demonstrating no blood flow proximal to the coils.

Case Presentation 2

A 66-year-old woman presented with vertigo and a right petroclival meningioma on MRI **(Fig. 14.6)**. At time of our initial consultation, her vertigo had resolved. However, she reported intermittent episodes of dizziness with a near-syncopal sensation, as well as fullness of the right ear. Evaluations by a cardiologist and an otologist were noncontributory.

We explained the natural history of meningiomas to her, and told her that the tumor was unlikely the culprit of her symptoms. We recommended a 3-month surveillance MRI, which was stable. Despite our recommendation for further monitoring, she expressed a strong desire for surgical intervention as the lesion was causing significant anxiety, which negatively impacted her quality of life.

In this asymptomatic patient with minimal brainstem compression, our goal for surgery was maximal partial resection without iatrogenic injury. We believed that the best approach to achieve this goal was a middle fossa craniotomy and anterior petrosectomy. At the end of surgery, our impression was that, if not all, at least most of the tumor had been removed. As the surgical goal was certainly achieved, the operation was terminated with little hesitation. The postoperative MRI, however, revealed a small residual at the caudal-most aspect of the tumor, between the IAC and the jugular tubercle **(Fig. 14.6c, d)**. Despite this, she was neurologically intact and the residual tumor has remained stable during follow-up.

Despite the tumor extension below the IAC, we knowingly chose not to perform a combined anterior–posterior petrosectomy to specifically and widely expose the caudal pole of the tumor. Adding the posterior petrosectomy would have significantly increased the complexity of surgery and lengthened the operative time. As the goal was set for maximal partial resection, our approach was uniquely designed to achieve this in the most expeditious way. The residual tumor will be carefully monitored in this woman nearing her eighth decade in life, and will be treated accordingly if it progresses.

■ Focusing on the Goals During Execution of Plans

During the treatment phase, the goals also help us get out of trouble. Execution of complicated plans seldom proceeds without

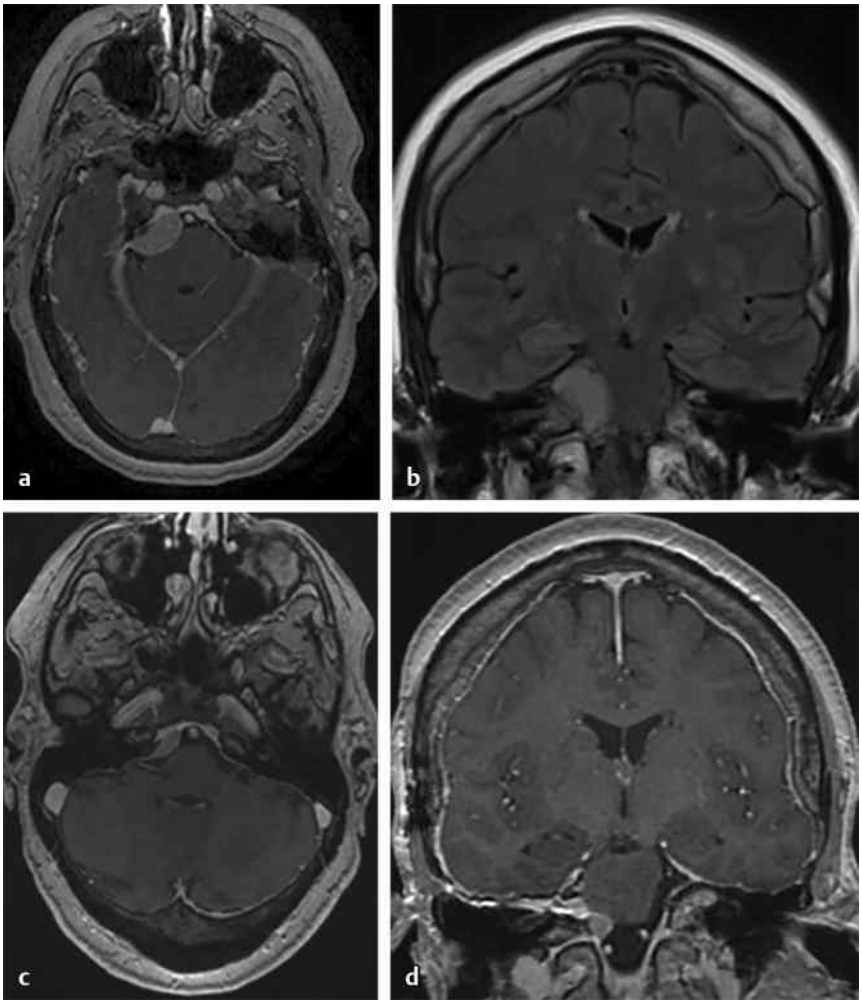


Fig. 14.6 Images of Case 2. (a) Preoperative axial MRI brain demonstrating an enhancing petroclival meningioma measuring 2.6 × 1.2 × 2.6 cm which extended from just below the internal auditory canal (IAC) to the level of the tentorium. **(b)** Coronal FLAIR MRI demonstrating the same mass. The inferior pole of the tumor lies just below the IAC and above the jugular tubercle. **(c)** Postoperative axial and **(d)** coronal MRI brain demonstrating a small residual tumor measuring 1.4 × 0.6 × 2.0 cm that extended just below the IAC and just beyond the reach obtained by an anterior petrosectomy. This lesion has been stable over the course of 17 months and, most importantly, the patient remains neurologically intact.

unexpected deviations, and keeping the goals in mind always help you find the right course out of the quagmire.

Case Presentation 3

A 67-year-old man presented with decreased fine motor functions of both hands, mild difficulty with ambulation, and intermittent dizziness. An MRI demonstrated a large petroclival meningioma measuring 5.9 × 5.5 × 3.9 cm (**Fig. 14.7**). On neurological examination, he had a partial left ptosis and 4+/5 strength in the right leg..

Our goal was set to be (1) brainstem decompression and (2) maximal resection without iatrogenic injury. Accordingly, the plan was to perform a two-stage surgical resection of this tumor, first with a FTOZ craniotomy and then an anterior petrosectomy. Prior to surgery, we obtained a cerebral angiogram to characterize the vascular anatomy, but the patient suffered a significant left-hemispheric stroke from this diagnostic procedure. The post-angio MRI demonstrated several areas of embolic infarcts, including the genu of the internal capsule and the left motor cortex (**Fig. 14.7c–e**).

Given the unexpected deviation in the plan, we changed the surgical approach to a single-staged operation to expedite his road to recovery. Believing that this would still achieve our goals and straighten our plans, we performed an FTOZ craniotomy for partial resection of the petroclival meningioma. Following surgery, the patient had a complete left ptosis and just trace movement of the right upper extremity and proximal right lower extremity. To further complicate matters, he suffered a pulmonary embolism following surgery, requiring anticoagulation.

The patient has shown significant recovery 8 months postsurgery. He has near-complete resolution of left ptosis, and is able to ambulate. His MRI showed tumor residual that has remained stable since surgery (**Fig. 14.7f**). Given his excellent recovery, we have abandoned plans for the second stage, and believe that it might do more harm than good.

■ Commentary

In the case presented by Dr. Nakaji and colleagues, the goal was clearly delineated upfront—brainstem decompression and maintenance of quality of life. That mindset helped determine the proper approach for their patient. Not all meningiomas need to be resected, and they prudently identified that the component of the tumor in the anterior fossa was simply a distraction, and that the main problem for the patient was the posterior fossa component. The design of the surgical plan was then focused only on the most straightforward path to eliminating this problem.

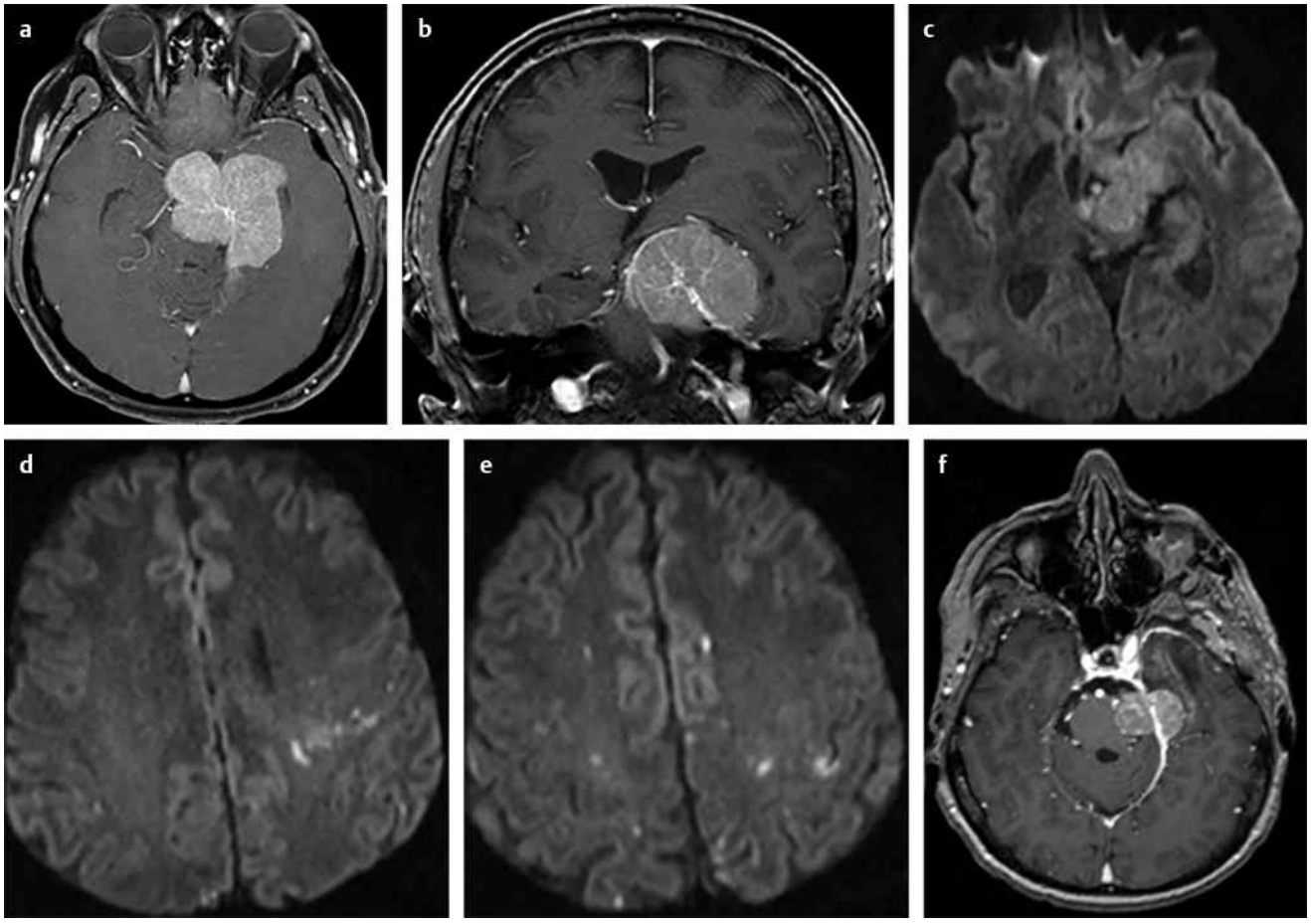


Fig. 14.7 Images of Case 3. Preoperative **(a)** axial and **(b)** coronal MRI brain demonstrating a 5.9 × 5.5 × 3.9 cm petroclival meningioma. The mass was causing significant compression of the brainstem and pushes the basilar artery contralaterally. **(c–e)** Diffusion-weighted imaging MRI postangiogram showed embolic infarcts on the left side of the brain including the genu of the internal capsule and the motor cortex. **(f)** Axial MRI brain following a frontotemporal orbitozygomatic craniotomy for subtotal resection showed residual tumor. The supratentorial portion measures 2.7 × 2.0 × 2.9 cm and the infratentorial portion measures 2.4 × 1.7 × 1.8 cm. This has remained stable over the course of 8 months. Most importantly, the patient has shown significant neurological improvement. The residual mass will be monitored with serial MRI scans.

Not all residual tumors are bad and not all meningiomas need to be removed. In fact, for the majority of meningioma cases which do not require emergent surgical intervention, we firmly believe in an upfront brief period of surveillance. We often obtain all the aforementioned additional imaging at 3 months in order to get to know the patient and family and also to assess for any growth or changes, both clinically and radiographically. At this point in time, we make the decision for continued surveillance or treatment based on growth and symptoms. Radiation treatment is an important option to consider as well. In addition to surgical resection, some patients are symptomatic from associated hydrocephalus, and shunting alone can improve quality of life without any need for surgical intervention. This is particularly important in older patients with slow-growing tumors. These non-resective strategies are also critical when planning a surgical approach because they are useful adjunctive options in cases where subtotal resection is achieved, planned, or otherwise.

Regarding the surgery described by Dr. Nakaji, we would have performed the operation with a few slightly different nuances. For positioning, although we have used the supine position before, we find the lateral position with the shoulder dropped slightly forward, and the head–neck angle opened up, to be more favorable. This position allows a better angle of attack no matter the size of the patient's neck/shoulder. In addition, the contralateral venous drainage is not compromised in this position and the patient's neck remains in a slightly more neutral position if he or she has mobility issues. For the dural opening, we favor a dural opening that is hinged on the sigmoid sinus. This allows early CSF drainage from the first, inferior cut in the dura as well as the possibility of protecting the flap from drying out during the case to allow possible primary closure. Of course, watertight closure is paramount and grafts are used as necessary. Because of the time and patient dissatisfaction with pericranial harvest, we prefer nonautologous grafts.

■ Conclusion

Over the years, we have been tempered in our surgical approach because of a general shift toward increased focus on quality-of-life goals, in addition to oncological and neurological ones. Although we regularly strive for complete tumor resection for oncological reasons, we always focus on what is best for the individual patient when considering all perspectives. Ultimately, our patients put their lives in our hands and we must do what is right for them.

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15 Petroclival Fissure

Jamie J. Van Gompel, Jeffrey R. Janus, Brian A. Neff, Joshua D. Hughes, and Jonathan Morris

Keywords: calcifying pseudoneoplasm of the neuraxis, chondrosarcoma, staged approach, "far medial"

■ Case Presentation

A 31-year-old woman presented to an outside facility with progressive neck pain. She stated she felt that her neck was "crumpled." She was evaluated and ultimately found to have a clival mass extending into her upper neck (**Fig. 15.1**).

At the outside institution, the patient underwent a right-sided suboccipital approach for biopsy, but the results were nondiagnostic. Days later, she underwent an occipitocervical fusion presumably to alleviate neck pain from cervical instability. As the first try was unsuccessful, a second biopsy was performed on the intradural portion of the tumor, but this resulted in significant left-sided weakness and other complications necessitating a percutaneous gastrostomy tube and a temporary tracheostomy for several months. After a long hospital course, she was followed by serial magnetic resonance imaging (MRI) scans. Because of slow growth of the mass and increasing neck pain, she presented at our institution. At this point, her left-sided weakness had improved, and despite spasticity, she was able to ambulate. Unfortunately, she had new deficits of progressive hypoglossal nerve weakness and a partial seventh nerve paresis.

Questions

1. What additional studies would help clarify what type of tumor this is?
2. Was a suboccipital approach the best way to biopsy the lesion?
3. What kind of further assessments should be performed after detecting tumor growth on follow-up?

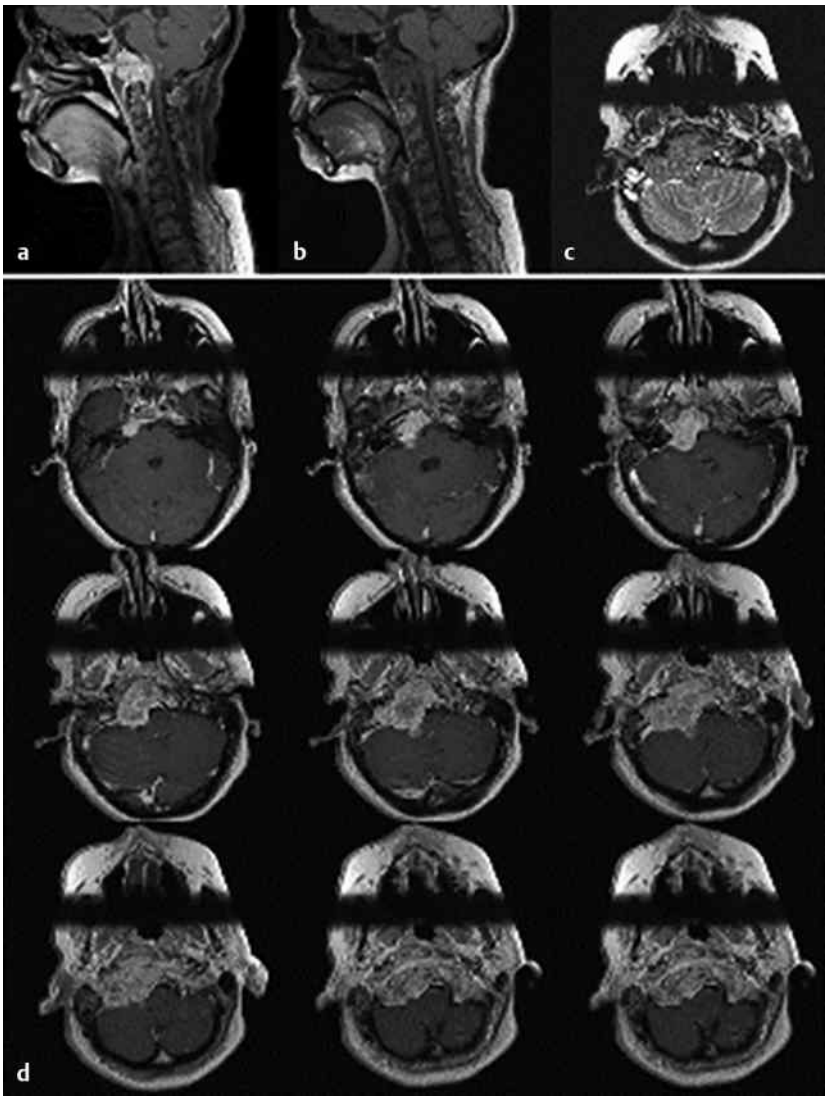

Fig. 15.1 Preoperative MRI. (a) T1 sagittal with and **(b)** without gadolinium demonstrating a contrast-enhancing clival mass. **(c)** Axial T2 and **(d)** T1 with gadolinium further demonstrating the extent of the mass at presentation.

■ Diagnosis and Assessment

As it appeared on the initial MRI, the differential diagnosis of the lesion includes chordoma, chondrosarcoma, and fibrous dysplasia. A biopsy was a reasonable first step in management, as the histopathological identity might significantly influence the course of action. However, the use of the suboccipital approach for the biopsy was suboptimal, as the majority of the mass was intraclival. Less invasive approaches, such as transnasal or transmastoid, might have been more effective. It is unclear why she underwent a cervical fusion since there was no investigation documenting any cervical instability. Presumably, stabilization was carried out to provide pain relief, which, unfortunately, it did not.

The second biopsy yielded the diagnosis of a "calcifying pseudoneoplasm of the neuraxis (CAPNON)," which is a rare, inflammatory disease. Repeat imaging demonstrated a large, slowly growing, calcifying pseudotumor (**Fig. 15.2**) causing medullary and brainstem compression. Importantly, there was now a T2-signal change at the upper portion of the mass, suggestive of cystic degeneration. The clinical significance of this change was unclear. Whereas in fibrous dysplasia, a disease far more common in this anatomical location, there is a risk of malignant degeneration into chondrosarcoma when such changes are seen, whether this transformation happens in CAPNON is unknown. The intense contrast enhancement of the tumor on MRI suggested the lesion was very vascular. Records from the prior operations were of little value for confirming this, as there was injury to the jugular foramen which generated significant bleeding of its own.

At our initial consultation, a baseline audiogram showed that her sensorineural hearing was normal, but cranial nerves IX through XII were dysfunctional on the right. A new computed tomography (CT) demonstrated a well-healed and fused craniocervical junction with no evidence of pseudoarthrosis. Therefore, cervical instability was unlikely to be the main cause of her worsening pain. A thorough literature search unearthed a case report about a symptomatic, calcified pseudotumor of the thoracic spine that resolved with indomethacin treatment. Since the patient's primary new complaint was neck pain, we saw no disadvantage to a trial of indomethacin. Because of the concern for malignant degeneration, we recommended a repeat biopsy of the clival portion of the mass via an endoscopic, transnasal approach. Given her prior experience and complications, she understandably declined yet another biopsy but agreed to undergo indomethacin treatment.

After 6 months of indomethacin, the patient's pain has improved. MRI revealed a stable mass and the lack of response to indomethacin treatment was in stark contrast to the thoracic calcified pseudotumor in the case study cited above. After an additional 6 months of indomethacin therapy, her urinary protein began to elevate, and she began to show signs of chronic renal insufficiency. The drug was stopped and a repeat MRI demonstrated progression in both the tumor and the cystic portion. At this point, we recommended aggressive resection.

Preoperative Assessment

Since there were hints suggesting a highly vascular tumor, an angiogram was performed in preparation for surgery. This showed a posterior skull base tumor eccentric to the right side with a vascular blush similar to a meningioma, and a small amount of arteriovenous shunting consistent with a highly vascular tumor (**Fig. 15.3**). Devascularization of tumor blood

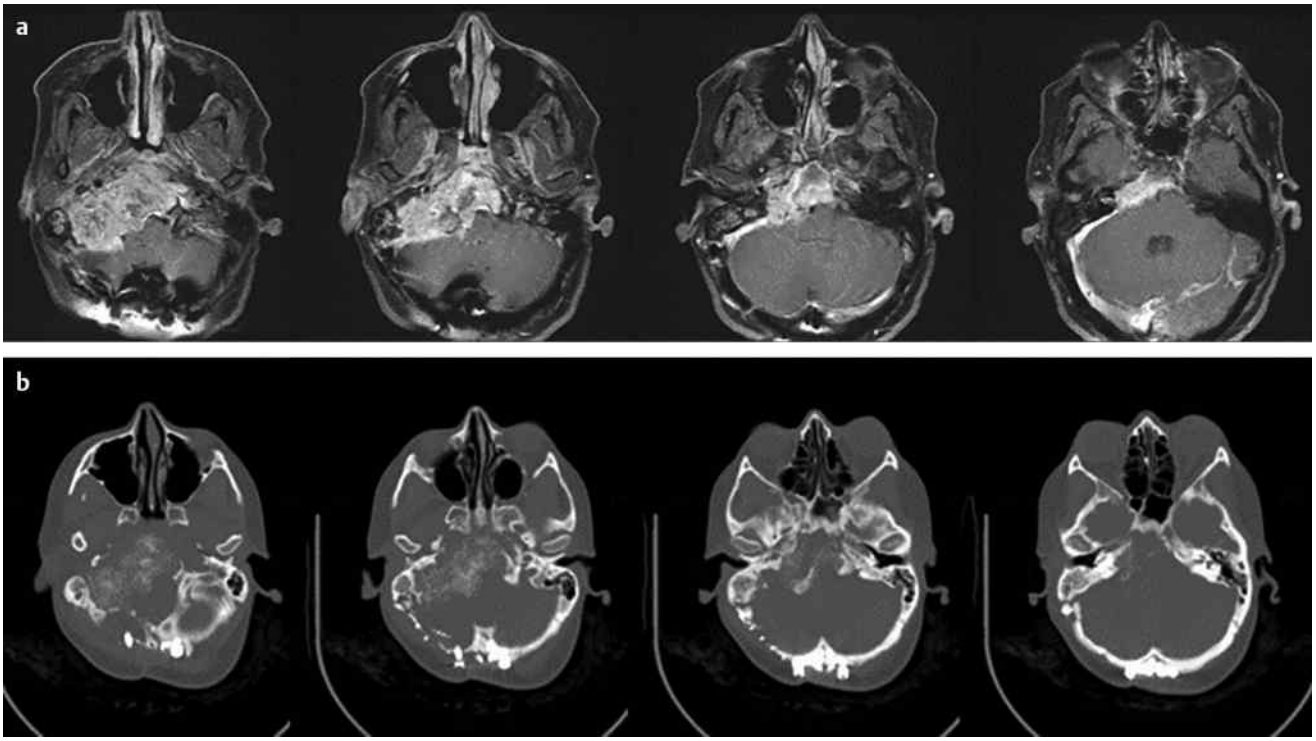

Fig. 15.2 Preoperative images. Serial aligned axial images: **(a)** MRI T1 with gadolinium and **(b)** noncontrast bone window CT images demonstrating imaging features of substantial contrast enhancement and bony consistency of the lesion.

supply was accomplished by embolizing both occipital arteries and both ascending pharyngeal arteries. The tumor blood supply from the right vertebral artery was also partially reduced by embolizing the posterior meningeal artery. Thoughts of further embolization through the right vertebral artery were abandoned as it could jeopardize the posterior inferior cerebellar artery. The embolization material used was 350 to 500 µm polyvinyl alcohol particles. It should be noted the jugular system on the right side showed long-term occlusion from the tumor and previous biopsy procedures.

In preparation for such a complex skull base procedure that may require uncommon approaches, we have the good fortune of an excellent radiology department with advanced segmentation abilities capable of three-dimensional printing of the tumor (**Fig. 15.4**). This improved surgeons' preoperative planning and communication with colleagues for multispecialty team cases. Furthermore, such three-dimensional visualization significantly helps in the planning of the crucial skull base reconstruction. Finally, after these tumors are removed, we typically provide the patients with a copy of the three-dimensional model to show them the tumor is gone, and this often has a significantly positive emotional impact.

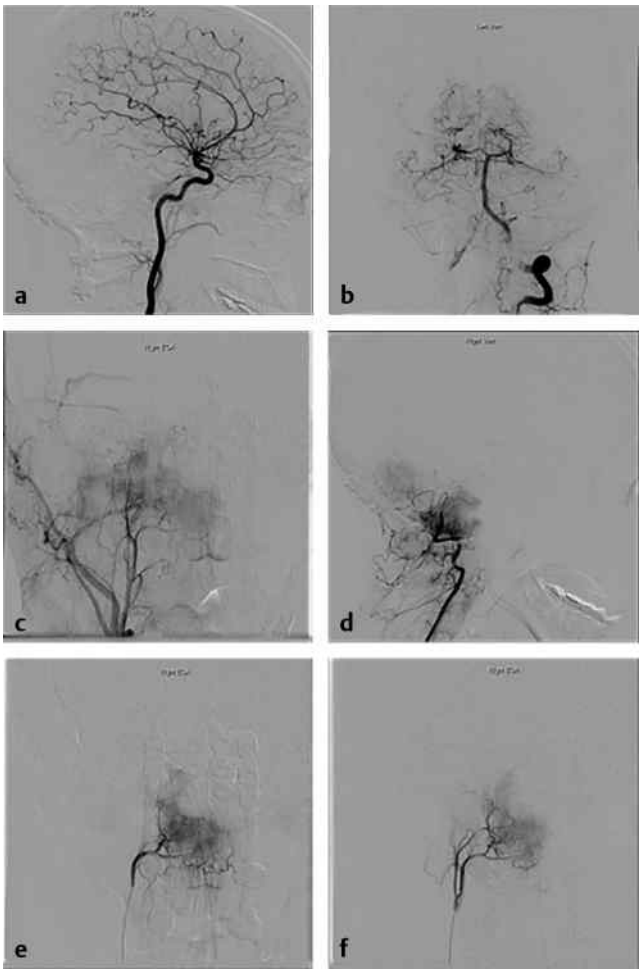

Fig. 15.3 Preoperative angiogram and embolization of the tumor. **(a)** Right internal carotid artery (ICA) injection demonstrating normal ICA without internal contribution to the tumor despite its proximity in the petrous bone. **(b)** Vertebral artery, **(c)** right external carotid, **(d)** right ascending pharyngeal, and feeding **(e)** right and **(f)** left occipital artery injections showing the tumor vascularity.

■ Anatomical and Therapeutic Considerations

While the need for resection was no longer up for debate, the question still remained if a complete resection was necessary. Although the lower cranial nerves on the right were not

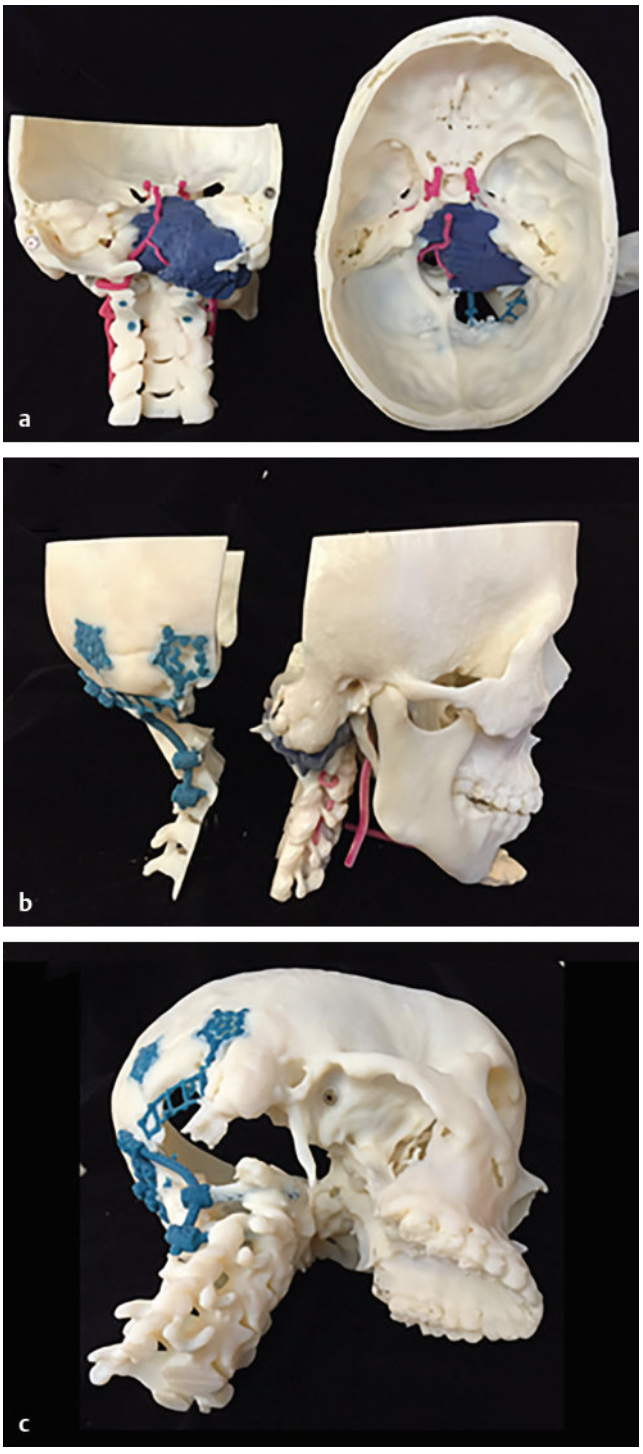

Fig. 15.4 Three-dimensional model of the tumor with **(a)** back-to-front and top-down view (tumor is in *blue*). **(b)** Lateral view showing the model has been severed to allow better analysis of the three-dimensional anatomy and **(c)** oblique view showing the skull base defect after the tumor has been removed.

functional, it was unclear whether this was caused by infiltration of the nerves or compression by the lesion. If it were the latter, there was a possibility that decompression alone could lead to some recovery. Operating under the assumption that complete resection was not necessary for this nonneoplastic disease process, we set our surgical goals for maximal subtotal resection without incurring additional neurological injury, and histopathological diagnosis to rule out malignant transformation.

Options for Approach

There was substantial tumor burden within the clivus and the petrous apex posterior, medial, and superior to the carotid. The jugular and hypoglossal foramen were completely encased with tumor; however, the cochlea and labyrinth were spared. We considered using the posterolateral corridor for the surgical approach. A transsigmoid approach would allow tumor resection through an opened jugular foramen. Although this would increase the risk of lower cranial nerve injury, this patient already had significant preoperative deficits in these nerves. One could also consider sacrificing hearing with a translabyrinthine approach or, even more aggressively, a transcochlear approach. In our patient, with practically normal hearing and facial function, these options were suboptimal.

Approach of Choice

In fact, it was difficult to come up with one approach that would completely remove the mass without destruction to healthy nerves. Our strategy for tumors like this is to break down the procedure into components and assess the best approach to access each of these locations. We decided that the best way to approach the cystic, degenerative portion of the tumor was to utilize the endonasal corridor in the first stage of a two-stage operation. Performing this first would give us the histopathological data to affect our surgical strategy for stage 2. The standard endoscopic transclival approach yields a trapezoid-shaped opening with the narrow end toward the foramen magnum. In order to reach the more lateral structures such as the jugular tubercle and foramen, the "far-medial" extension would be necessary, the term having been coined for the endoscopic technique to access the same areas reachable by the "far-lateral" approach in "open" surgery.

A second-stage surgery was planned at a separate setting via a transmastoid infralabyrinthine approach, taking advantage of the window created by the tumor between the jugular foramen and the internal acoustic canal (IAC). We would open the jugular bulb, but not transgress the back wall into the neural compartment containing the lower cranial nerves. Moreover, since the lesion was not malignant, we intended to preserve the lower cranial nerves with the hope that they would eventually recover function with decompression (**Fig. 15.5**).

Questions

1. What are the lateral limits of the "far-medial" extension in the endonasal endoscopic transclival approach?
2. How far apart would you separate the two stages of the surgical plan?
3. What nerves would you monitor during each stage of intervention?

■ Description of the Technique

Stage 1: Expanded endonasal transclival approach with far-medial extension.

The patient was placed in the supine position without a lumbar drain and a stereotactic CT angiography was utilized. A left nasoseptal flap was made and swung into a left maxillary antrostomy. The patient had prominent posterior inferior turbinates, which were removed as well as the adenoid. Through a wide sphenoidotomy, we identified the vidian canals bilaterally and skeletonized these back to the lacerum portion of the carotid. This exposure provided a large operative window to the clival portion of the tumor which had a firm capsule around it (see **The Three Approach Elements 1**).

Even with embolization, the lesion was extremely vascular and firm. It also had calcifications within it that could only be

The Three Approach Elements 1

Corridor: endonasal

Craniotomy: N/A

Modifiers: transclival/far-medial

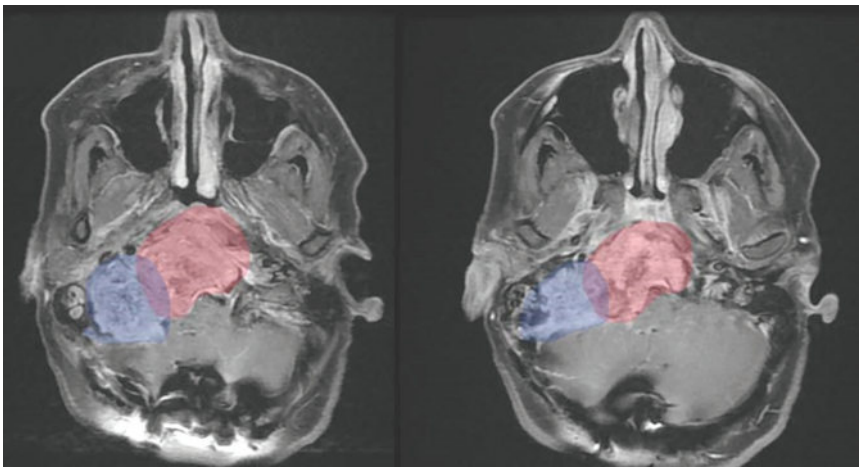

Fig. 15.5 Two-stage operative plan. The approximate surgical targets of the two operative approaches are shown here. The endoscopic, endonasal transclival approach with far-medial extension would remove the area in red, depending on intraoperative findings. The transmastoid, infralabyrinthine approach was planned as stage 2 to remove the areas in blue as far as the surgical exposure allowed. The borders between the two are "imaginary," and shown here only to conceptualize the surgical design.

removed piecemeal (**Fig. 15.6a**). Intraoperative frozen section diagnosis was a spindle cell neoplasm favoring meningioma; however, the final pathology was CAPNON. Specifically, we biopsied the area that had cystic degeneration separately, and this portion showed no histological differences from the rest of the CAPNON. We worked with 0-, 30-, and 45-degree endoscopes and ultimately were able to resect tumor until we visualized and stimulated the sixth cranial nerve within the tumor (**Fig. 15.6b**). We separated the tumor from the dura which was incompetent in several areas. The intradural tumor was removed without difficulty. The tumor itself had done much of the exposure for us as the bone of the hypoglossal canal and the occipital condyle was replaced by the lesion. This exposure was more lateral than the typical endoscopic transclival procedure.

We also identified the hypoglossal nerve, and even though it did not stimulate, we removed the tumor away from this. Next, we worked to the top of the odontoid and removed the tumor and capsule in this area as well. At this point, we had achieved as much as could be expected through the endoscopic approach. Despite preoperative embolization, the patient required 2 units of blood during the case. At the conclusion of the resection, we put collagen substitute intradurally and placed a fascia lata graft over the dura. Afterward, the nasoseptal flap was rotated into position. This resection resulted in approximately 40% of the tumor being removed.

Stage 2: Transmastoid, infralabyrinthine approach with endoscopic assistance.

This stage was performed approximately 3 months after the endoscopic procedure to allow for maximal recovery and reduction of risk of complications. The patient was placed in the lateral decubitus position. Stealth CT angiography was once again used for orientation. With the mastoid exposure, we used a typical, small incision as we did not want to expose the occipital cervical instrumentation. We exposed the mastoid and prior suboccipital bone work, and removed some of the instrumentation anteriorly that had been used to affix the prior suboccipital craniotomy near the mastoid (see **The Three Approach Elements 2**).

The Three Approach Elements 2

Corridor: presigmoid (transsigmoid)

Craniotomy: transmastoid

Modifiers: infralabyrinthine petrosectomy

A tympanomastoidectomy was performed leaving the tympanic membrane untouched. The sigmoid sinus was decompressed completely. Next, the jugular bulb was identified and decompressed as well, thus exposing the tumor which filled the jugular bulb (**Fig. 15.6c**). Next, we skeletonized the mastoid segment of the facial nerve as well as the labyrinth. We opened the jugular bulb, which was filled with tumor, but we kept the back wall intact in order to stay extradural and to minimize the chance of lower cranial nerve injury. We internally debulked the tumor while identifying the posterior fossa dura, and we worked through the corridor between the jugular bulb and labyrinth (infralabyrinthine) to remove sequentially deeper tumor extending into the posterior petrous apex. With the aid of neuromonitoring, we were able to stimulate and identify the facial nerve within the proximal portion of the IAC nearest to the porus acusticus and the lower cranial nerves extradurally (**Fig. 15.6d**). After tumor removal, we were able to stimulate cranial nerves IX, X, and XI at 1 2 mA. Working further

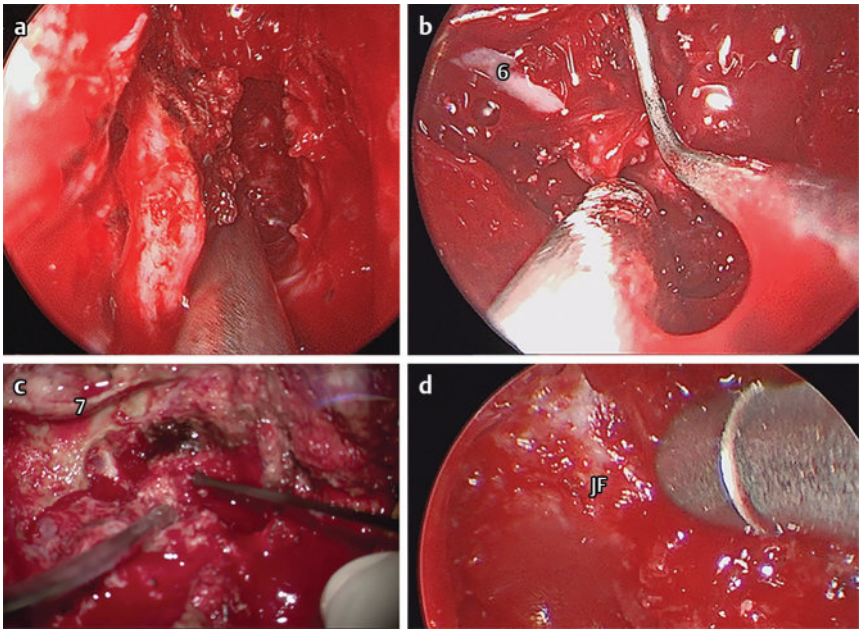

Fig. 15.6 Intraoperative images. **(a)** endonasal endoscopic exposure of the tumor demonstrating the calcified, granular, and vascular tumor. **(b)** After initial removal of the tumor, within it was found the sixth cranial nerve (6) and the tumor was removed from around this. **(c)** Stage 2 transmastoid, infralabyrinthine approach demonstrating the corridor after the mastoid had been removed (7, seventh nerve in the mastoid). **(d)** After initial removal of the tumor with the microscope, the endoscope was advanced into the cavity between the IAC and the lower cranial nerves. Here, we were removing tumor from the jugular foramen (JF).

inferior to these nerves, spontaneous firing from cranial nerve XII was detected. We dissected and completely skeletonized cranial nerve XII within the hypoglossal canal. We then worked inferiorly from this, resecting tumor down to the occipital condyle. Here, posteriorly, we found the fat over the vertebral artery and resected the tumor from this region.

We then worked anteriorly to resect as much tumor as possible. It should be noted, that preserving anatomically intact cranial nerves probably limited our resection, but, despite preoperative partial paresis of these nerves, we felt that decompression with neural preservation was the goal. We stuck with this philosophy especially given the intraoperative response of the lower cranial nerves. Ultimately, we worked through our prior resection cavity medially and identified the fascia lata that was left behind from the transnasal procedure. Here, there were two heavily ossified plateaus of bone, which were adherent to the dura and protected it. We had previously encountered these during the endonasal procedure. We left them intact, as they seemed to represent recreation and ossification of the clivus rather than recurrent CAPNON.

At that point, we had an aggressive, subtotal resection. Inspecting the cavity with a 45- and 60-degree endoscope mounted on a Mitaka arm, we found more tumor deep to the jugular foramen looking superiorly. Using curved curettes, more tumor was removed. We left a small portion of tumor over the superior aspect of the petrous apex overlying the IAC, as well as a small portion over the carotid artery. Reinspecting with a 60-degree endoscope, we were satisfied with our overall resection. Abdominal fat graft was used to fill the defect medially after it was thoroughly irrigated. A piece of pressed temporalis fascia was placed over the antral opening to the epitympanum and middle ear. Tisseel was placed on top of this. The fat was used to obliterate the mastoidectomy and mastoid tip area. The incision was then closed in layers.

Surgical Pearls

1. In both approaches, the utilization of endoscopic assistance reduced the morbidity to the patient and enhanced visualization and the degree of resection.
2. The two stages of the operation were separated by 3 months in this patient. The time gap needs to be considered in relation with the preoperative growth rate of the lesion. Since the lesion in this patient evolved very slowly, we believed that a relatively long time gap of 3 months was in her best interest. While our intention was to do a third stage to address the small amount of tumor left behind on the petrous carotid, we are currently monitoring this hoping for spontaneous regression.
3. Preoperative embolization resulted in a much more manageable lesion during surgery. Furthermore, when a mass of this size involves the jugular foramen, angiographic documentation of the functional flow and venous anatomy is critical to avoid further morbidity. This is simply not feasible with noninvasive techniques such as MR venography.

Aftercare

At the 15-month follow-up, the patient stated her headaches have improved substantially since partial resection (> 90%) of her CAPNON. We discussed a third operation to remove the portion adjacent to the petrous carotid artery via a middle fossa craniotomy combined with an anterior petrosectomy; however, the patient elected to watch the residual since her symptoms have improved. Interestingly, on postoperative CT scans, it appeared that the inferior portion of the lesion is calcifying or hardening (**Fig. 15.7**). These changes indicate probable bony regrowth instead of recurrent inflammatory pseudotumor.

Very little is known about best practices for treating this inflammatory process. Although most case reports involve lesions which were resectable completely with minimal morbidity, our patient had an extensive lesion in a relatively difficult area to access surrounded by many critical structures. A complete resection would have led to significant cranial nerve morbidity. This was not warranted since the pathology was nonmalignant, and the patient had already suffered enough from the biopsies. Curiously, as it appears that no recurrence has been detected in our patient, the real question remains whether complete resection is ever necessary and whether immune modulation will result in eventual resolution.

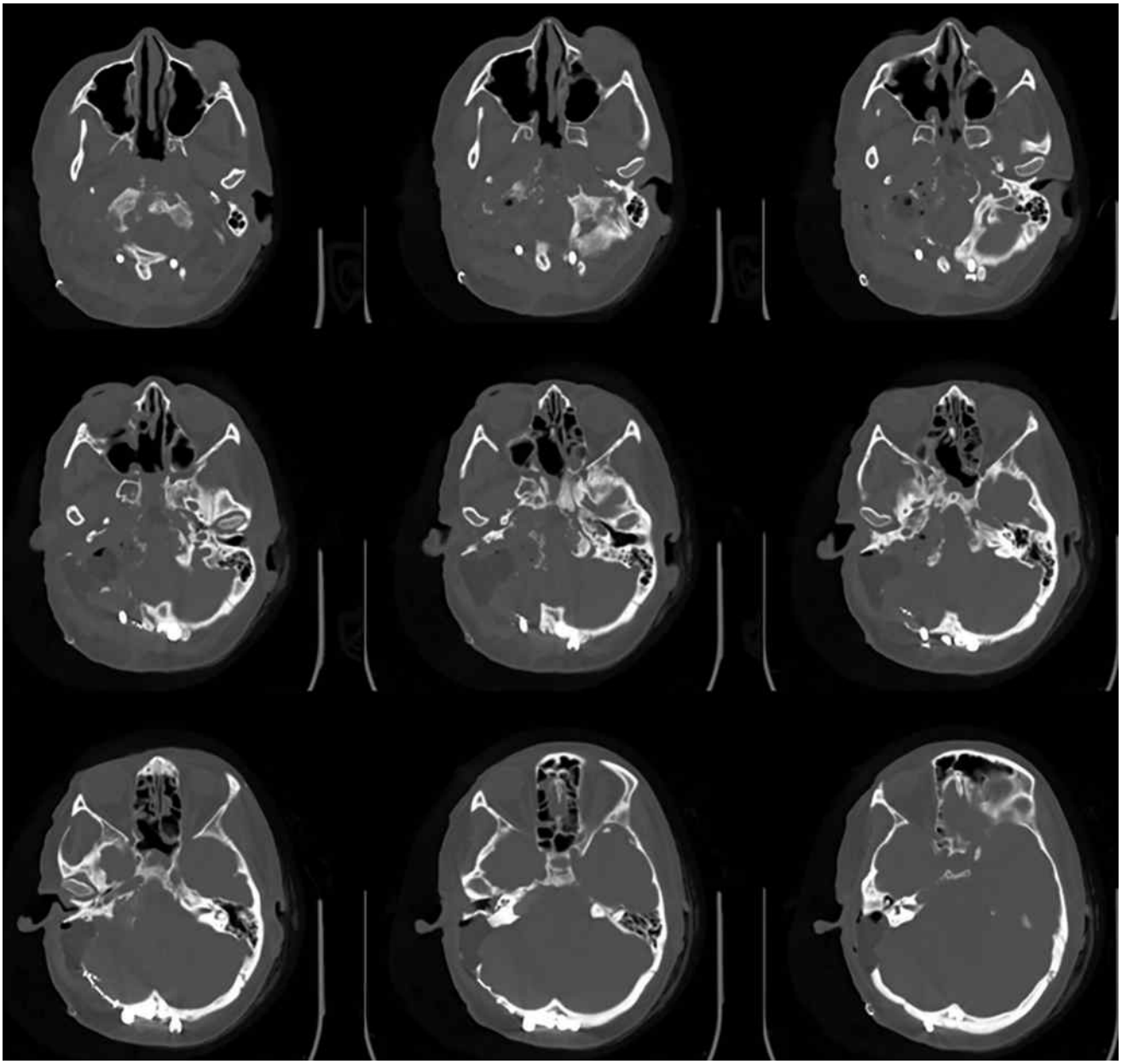

Fig. 15.7 Postoperative images. Serial axial images at 3 months showing the final resulting resection. There was a small residual at the petrous apex.

Perspective

Maria Koutourousiou, Paul A. Gardner, Carl H. Snyderman, and Eric W. Wang

■ Introduction

Bone tumors of the petroclival fissure are extremely challenging because of their deep location, extensive involvement of multiple cranial fossae and foramina, and compression, or even invasion, of the brainstem. Questionable results of radiation in controlling chordomas and chondrosarcomas prompt surgeons to be aggressive but radical resections carry a substantial risk. For transcranial surgery, the cranial nerves are interposed between the surgeon and these deep-seated tumors, making the risk of cranial morbidity very high. The endoscopic endonasal approach (EEA) has been used since the beginning of the 21st century for clival tumors, and it allows direct access to the clivus without brain retraction and nerve manipulations. The major limitation of the endoscopic endonasal transclival approach is the lateral extension of the lesions behind the internal carotid artery (ICA) and beyond the petroclival fissure, but techniques are evolving to overcome this limitation.

■ Endoscopic Endonasal "Far-Medial" Approach—Advantages and Limitations

The endoscopic endonasal transcondylar and transjugular tubercle approach (or "far-medial" approach) provides a unique surgical corridor from the inferior third of the clivus to the ventrolateral surface of the ponto- and cervicomedullary junctions. Just as a dorsolateral condyle resection (i.e., transcondylar approach in "open" surgery) added to the lateral suboccipital approach allows for a more medially directed trajectory, a ventromedial condyle resection added to the endoscopic endonasal inferior transclival approach allows for a more laterally directed corridor. With the addition of the "far-medial" approach, the transclival EEA becomes an option for midline clival lesions even if the tumor extends lateral to the petroclival junction to the jugular foramen, hypoglossal canal, and/or occipital condyle. The medial border of the cranial nerve foramina and the ICAs represents the lateral limitations of this approach (**Fig. 15.8**). Access can be extended behind the ICA by "skeletonizing" it (removing the overlying bone) and carefully retracting the artery. Access to the medial petrous apex can be achieved through the "Gardner's triangle," defined by the abducens nerve, the paraclival ICA, and the foramen lacerum (**Fig. 15.9**).

Advantages

When a lesion is located anterior to the brainstem (like a clival/petroclival bone tumor), distortion and posterior displacement of the brainstem and cranial nerves make it difficult for

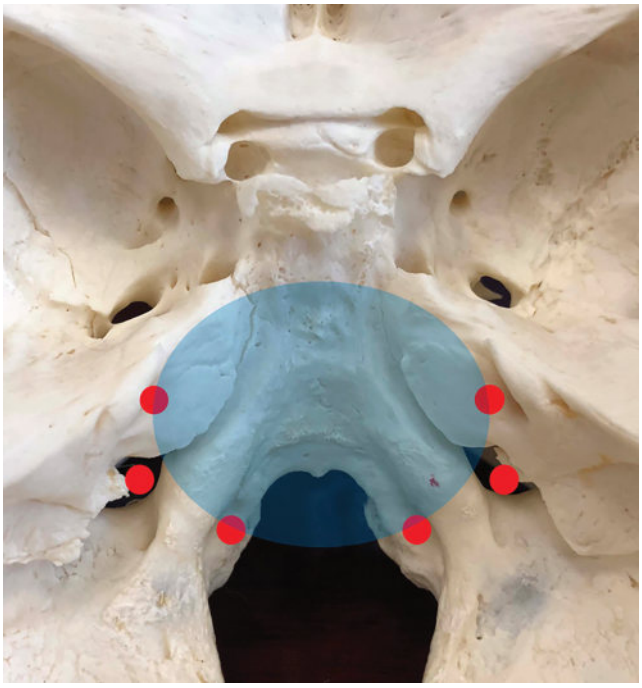

Fig. 15.8 Skull base region accessible via the endoscopic endonasal approach. The internal acoustic canal, jugular foramen, and hypoglossal canal are labelled with red dots on both sides. The blue area drawn between the cranial nerves represents the central skull base which is accessible through the endoscopic endonasal approach in the posterior fossa.

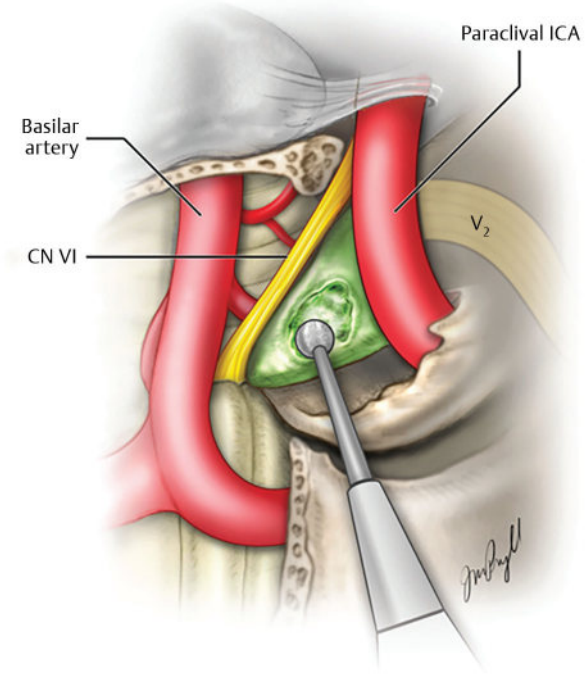

Fig. 15.9 The Gardner's triangle. Access to the medial petrous apex can be gained by carefully drilling behind the skeletonized paraclival internal carotid artery (ICA), through the entry known as "Gardner's triangle" (shaded in *green*). It is bordered by the paraclival ICA, the abducens nerve, and the line drawn across the foramen lacerum.

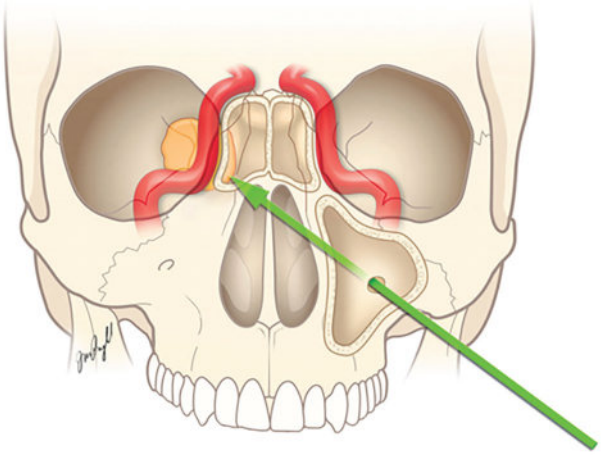

Fig. 15.10 The contralateral transmaxillary approach. This approach is designed to extend the lateral reach of the endoscopic endonasal approach toward the petrous apex. Through a contralateral maxillary antrostomy, the trajectory of the approach is deep to the paraclival internal carotid artery, and nearly parallel with the horizontal segment of the artery.

posterolateral approaches, since the lateral-to-medial trajectory of these approaches requires manipulation of the neurovascular structures within the narrow surgical corridor to reach the ventral midline region. The endoscopic endonasal transclival "far-medial' approach allows access to clival, petroclival, and foramen magnum lesions via a medial-to-lateral trajectory, providing a direct, unobstructed route to the lesion. With the endoscope, there are wider viewing angles, better illumination, and the ability to look around corners. Furthermore, in contrast to most lateral open approaches, the "far-medial" approach does not require additional openings in the dura of the cranial base other than those immediately involved with tumor, and there is no manipulation of the vertebral artery. Provided the approach is performed by experienced surgeons, the absence of brain retraction and manipulation of nerves and vessels result in less risk and a faster recovery period.

Contralateral Transmaxillary Approach (CTM)

This recently developed approach helps to improve the lateral access for midline approaches like the EEA. An anterior maxillary antrostomy (Caldwell–Luc) is performed on the contralateral side from the petroclival involvement. This corridor is nearly parallel to the course of the horizontal petrous ICA and therefore allows a more lateral-to-lateral (i.e., side-to-side) trajectory beyond the petroclival junction to the IAC and jugular foramen (**Fig. 15.10**). This is typically used in conjunction with an endonasal approach with dissecting instruments, suctions, and drills introduced via the contralateral transmaxillary (CTM) corridor. Exact limits, applications, and limitations of this approach have yet to be defined, but it holds great promise as an adjunct for a very challenging area to access.

Limitations

Extreme Lateral Tumor Location/Extension

Although the aforementioned new techniques have expanded the indications of EEA to lesions beyond the midline, tumors originating or extending lateral to the petroclival fissure remain challenging to reach. Even with angled endoscopes, lateral tumor extensions in the petrous bone are hard to see, and ill-advised "blind dissection" would increase the risk of complications and unintended tumor residual.

In addition, increasing lateral access requires complete skeletonization of the paraclival ICA for its mobilization and retraction. Although adding the CTM may reduce the need for this, the steep learning curve for the CTM may limit its common application.

For these reasons, many tumors located in the petrous apex, including the IAC and jugular foramen, are ill-suited for the EEA and an open transcranial approach is preferred. Tumors which *originate* lateral to the cranial nerves are contraindicated for EEA, as they displace critical structures medially, thus negating the key advantage of a medial approach. Finally, for giant clival/petroclival tumors with extreme lateral extension, there is often not a single approach suitable to reach the entire lesion, and a combination of EEA and open surgery is indicated.

Surgeons' Experience

Endoscopic endonasal surgery requires an experienced, multidisciplinary team and advanced equipment (angled-endoscopes, endoscopic-adapted dissection, coagulation and tumor ablation instruments) for safe results. Before attempting any expanded EEA, extensive training and experience are mandatory. Although there is no consensus on the exact number of surgeries necessary to achieve an acceptable level of expertise, it is critical that surgeons analyze their own team's experience and outcomes, before selecting a complex EEA for a patient.

Surgical Complications

One of the main criticisms of using EEA for extended skull base lesions is the high rate of postoperative cerebrospinal fluid (CSF) leak, and this is especially true for intradural tumors of the posterior fossa. Of course, if an osseous tumor is encased in bone without dural involvement, CSF leak is seldom a concern. However, some aggressive chordomas can invade the dura without appearing to do so on imaging studies. Multilayered reconstruction techniques using a vascularized nasoseptal flap and fat graft have significantly lowered the CSF leak rate, but these too have limitations. There is a clear correlation between obesity and CSF leak, and therefore, obese patients, and especially those who have a critical need for continuous positive airway pressure, are poorly suited for EEA. Similarly, patients with history of multiple prior endonasal surgery, who no longer have vascularized reconstruction options, should be advised to consider other options.

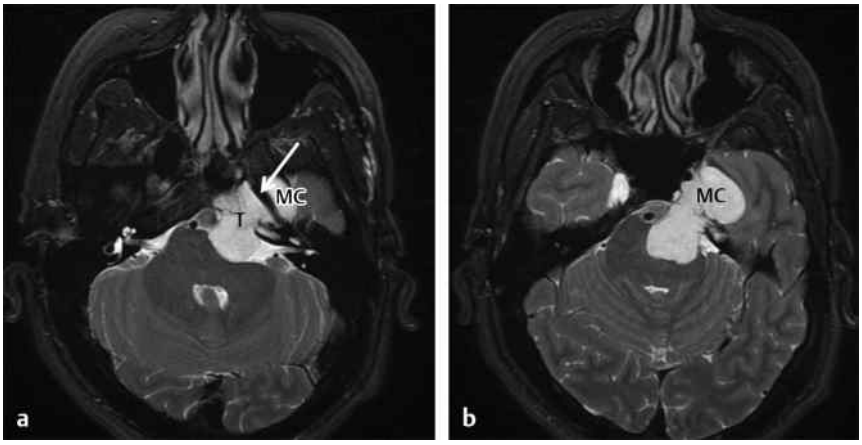

Fig. 15.11 Preoperative MRI. (a) Axial T2-weighted MRI images showing a large petroclival tumor (T) based around the petroclival fissure; **(b)** the tumor extended into Meckel's cave. *Arrow,* ICA; MC, Meckel's cave; T, tumor.

■ Case Presentation

A 53-year-old woman presented with diplopia from a left abducens nerve palsy. Preoperative images showed a large, T2-hyperintense mass centered on the left petroclival fissure (**Fig. 15.11**). The suspected diagnosis was a petroclival chondrosarcoma.

■ Anatomical and Therapeutic Considerations

For clival tumors with centers close to the midline, without lateral extension beyond the level of the IAC, jugular foramen, or hypoglossal canal, we prefer the transclival EEA, combined with the "far-medial" approach when necessary. Tumors centered lateral to the skull base foramina/cranial nerves are better suited for various lateral approaches, and the best option is dependent on the specific tumor's position on the clivus. For the upper clivus (i.e., sphenocavernous or tumors lateral to the optic nerves and cavernous sinuses), the preferred approach would be a pterional or frontotemporal orbitozygomatic approach. At the midclivus beyond the level of the IAC (petrous ridge), the transpetrous approach is used. The far-lateral or transcondylar approach is employed when the tumor is lateral to the jugular foramen or hypoglossal canal. As mentioned above, combining approaches may be necessary for giant tumors that extend beyond the reach of any singular approach, and at our institution, these combined approaches are always staged.

In the female patient described above, the surgical goal was a complete resection. The tumor originated medial to the cranial neural foramina and was therefore suitable for EEA. However, the extension into Meckel's cave would require a separate opening lateral to the ICA, even as the majority of the tumor can be resected via the corridor medial to it. The plan was to skeletonize the ICA and release its attachment at the foramen lacerum, thus allowing access to the petrous apex and petroclival fissure/synchondrosis to achieve our goal.

■ Description of the Technique

The patient was placed under general endotracheal anesthesia, supine on the operating table in three-pin Mayfield head fixation with slight head turn to the right. Electrodes for the neuromonitoring of the mid and lower cranial nerves (VI–XII) were placed. Image guidance was registered. The midface and abdomen were prepped and draped. Since invasion of the dura was considered highly possible, the vascularized nasoseptal flap was harvested at the beginning stages of the surgery.

The exposure started with wide bilateral sphenoidotomies, extensive maxillary antrostomy, and a transpterygoid approach on the side of petrous involvement. The posterior edge of the nasal septum was resected. Mucosa and fascia of the nasopharynx were then removed or released with monopolar cautery from the bottom of the sphenoid rostrum to the foramen magnum between the eustachian tubes. Once the clivus was entirely exposed, it was drilled from the sella to foramen magnum.

Next, the paraclival internal carotid arteries from the cavernous sinus all the way down to the foramen lacerum were skeletonized. The vidian nerve on the left was identified as a useful landmark. It was followed posteriorly to locate the foramen lacerum. The vidian nerve was then sacrificed for full exposure, since the dissection at the foramen lacerum was necessary to access the petroclival fissure. The bone overlying the left Meckel's cave was also removed, allowing further lateral mobilization of the ICA and access to tumor within Meckel's cave.

At this point, the tumor can be removed. Soft tumors such as chordomas or chondrosarcomas can be removed with suction and stimulating dissectors to localize cranial nerves. The abducens nerve was identified at the level of the Dorello's canal with the use of neurostimulator. Dissection of the nerve was carried from an inferior-to-superior trajectory, as petroclival bony tumors typically displace it superiorly.

Neurostimulation and image guidance were used throughout the tumor resection. A combination of 0-degree and angled endoscopes were used to fully explore the surgical field and

identify potential hidden tumor behind the left ICA (**Fig. 15.12**). Gentle packing with hemostatic agents and copious warm water irrigation were used to control venous bleeding from the basilar plexus. The tumor had intradural extension and CSF was encountered from the petroclival dural defect (**Fig. 15.12c, d**). In this case, after resection of the intradural tumor, care was taken to prevent surgical debris from entering the subarachnoid space.

For reconstruction, collagen matrix was used as an inlay, followed by fascia lata (or allograft), reinforced with autologous fat graft. The entire clival/paraclival area was covered with the vascularized nasoseptal flap. For large posterior fossa dural defects such as this one, packing is left in place for 6 to 7 days. Postoperative imaging showed a complete resection (**Fig. 15.13**), and the histopathological diagnosis was a chondrosarcoma.

Commentary

In the first part of this chapter, the authors discussed an extremely rare and complex case of a CAPNON. Given the large size, extreme lateral extension, and continuous growth of the lesion, there was no single approach which would be adequate to remove it without significant morbidity. A staged surgical resection would have been our choice as well.

Employing an EEA "far-medial" transclival approach, combined with a CTM, would be a good option for the first stage. Through a medial approach, there is less risk to the lower cranial nerves, while a generous debulking of the tumor, pathological confirmation of the lesion, and decompression of the brainstem can be achieved. However, it is critical to highlight that

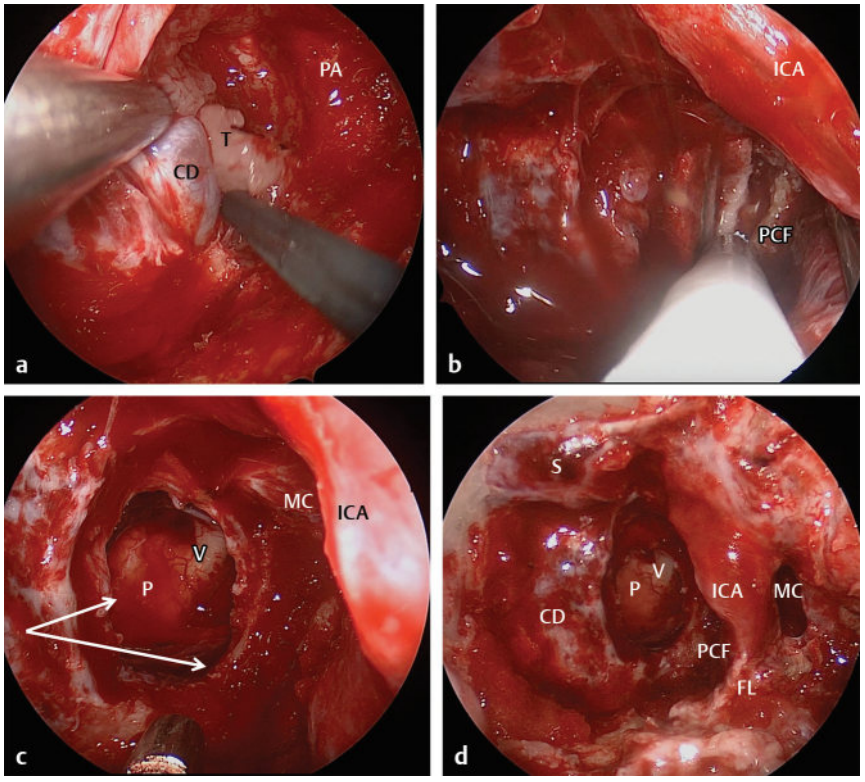

Fig. 15.12 Intraoperative endoscopic views. (a) The pale chondrosarcoma (T) was dissected from the lateral clival dura (CD) at the petroclival junction. PA, petrous apex. **(b)** Working behind the skeletonized internal carotid artery (ICA). The Sonopet ultrasonic bone curette was used to resect the petrous bone at the left petroclival fissure (PCF), which was involved with tumor. **(c, d)** The view following complete resection of a petroclival chondrosarcoma involving the lateral petroclival dura, petrous apex, petroclival fissure, and Meckel's cave (MC). *Arrows*, petroclival dural defect; CD, clival dura; FL, foramen lacerum; ICA, internal carotid artery; P, pons; PCF, petroclival fissure; S, sella; V, root of trigeminal nerve.

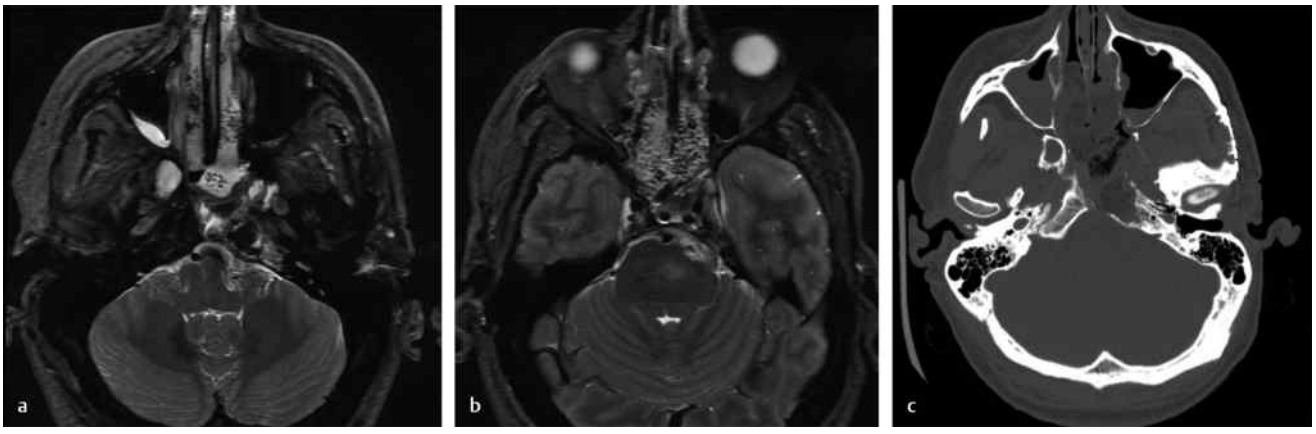

Fig. 15.13 Postoperative imaging studies. (a, b) Axial, T2-weighted MRI images showing complete resection of the large petroclival chondrosarcoma with only residual blood products in the tumor cavity. **(c)** Postoperative CT showing total resection of tumor and involved bone from the left petroclival region.

the complex endoscopic endonasal operation on the CAPNON patient was performed by an experienced team of surgeons. Surgeons who do not have the appropriate training and experience to perform EEAs for extensive skull base lesions should employ the approach with which they feel most comfortable.

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16 Foramen Magnum

Da Li, Huan Li, Zhen Wu, and Jun-Ting Zhang

Keywords: meningioma, occipital condyle, vertebral artery, hypoglossal canal, far-lateral approach, endonasal transclival approach

■ Case Presentation

A 48-year-old previously healthy man presented to a local hospital with progressive numbness in his extremities for 2 years. He also had weakness in the right arm and leg, which progressed over the last year, and he started to experience numbness in the upper chest. He denied having headaches, nausea, or problems with swallowing or phonation. On direct questioning, he admitted to having paresthesia in all four extremities. On neurological examination, he was found to have an abnormal gait, and was hemiparetic on the right and numb in the T 4–6 distribution.

During his evaluation, a noncontrast computed tomography (CT) scan revealed a mass, isodense to the brain, in the lower clivus without calcification. A subsequent magnetic resonance imaging (MRI) scan with contrast enhancement showed a foramen magnum lesion ventral to brainstem, compressing it posteriorly (**Fig. 16.1**).

Questions

1. Other than a meningioma, what should be considered in this location?
2. Is radiosurgery or surgery better for this patient? Why?
3. If surgery is considered, what preoperative tests are necessary to complete the work-up?

■ Diagnosis and Assessment

The MRI scans on T1- and T2-weighted images (**Fig. 16.1**) showed an isointense, foramen magnum lesion, ventroinferior to both vertebral arteries without obvious encasement of surrounding vessels. The contrasted MRI scan showed homogenous enhancement and the wide dural attachment of the lesion from the lower clivus to the level of C2. The CT scan was useful to assess the bone anatomy and the degree of calcification of the lesion (**Fig. 16.1e**). The differential diagnosis for a lesion in this area included inflammatory granuloma, neuroma, dermoid cyst, epidermoid cyst, neurenteric cyst, and chordoma. The dural tail on the contrasted MRI heavily favors the diagnosis of a meningioma.

In setting the treatment goals for this patient, it is important to note that his symptoms were progressing, and it was unclear how quickly he would deteriorate in the near future. In fact, his ventricles were mildly dilated, indicative of impending hydrocephalus. Clearly, expedient decompression of the brainstem was a necessity. As there was little doubt regarding the histopathological diagnosis, biopsy would be of little use. Observation and radiosurgery would be equally irrational, the latter mainly because of its inability to decompress the brainstem. Furthermore, because there was no space between the tumor and the brainstem, radiosurgery to this large tumor may in fact harm the very structure (i.e., brainstem) that it should protect.

Taking all this into account in this otherwise healthy patient, surgical resection was selected as the best treatment, with the goal of maximal removal without harming the brainstem and cranial nerves. Depending on intraoperative findings, if a total removal can be accomplished without harming the patient, it could potentially be curative. Lower cranial nerve function should be assessed in detail prior to surgery to determine preoperative status.

■ Anatomical and Therapeutic Considerations

For anatomical considerations related to meningiomas, the foramen magnum "region" has been defined by Bruneau and George with these boundaries: anteriorly, from the lower third of the clivus to the upper margin of the body of C2; laterally, from the jugular tubercle to the upper aspect of the C2 lamina; and posteriorly, from the anterior edge of the squamous occipital bone to the C2 spinous process. Before sorting through the surgical options available for this region, highlighting some anatomical nuances would be helpful.

The opening of foramen magnum is oval-shaped, and the posterior part is wider than anterior part, which is just rostral to odontoid process. The occipital condyle is located lateral to the anterior half of the foramen magnum on each side, and the alar ligament from the odontoid process attaches to the

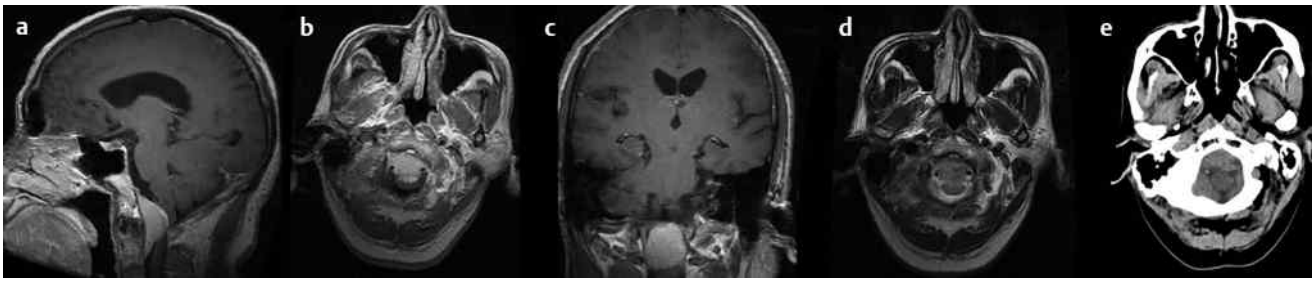


Fig. 16.1 Preoperative images. (a–d) MRI and **(e)** CT images showed a foramen magnum meningioma caudal to both vertebral arteries. The T2-weighted image in **(d)** showed that there is no subarachnoid space between the lesion and the brainstem.

anteromedial portion of it. The condylar fossa is located on the extracranial surface, directly behind the condyle (**Fig. 16.2**). The posterior emissary vein, traveling through the posterior condylar canal, empties into this fossa and it connects the vertebral venous plexus with the sigmoid sinus.

The most important artery of the region is the vertebral artery. The V3 segment extends from the C2 transverse process to the foramen magnum dura. It courses from the traverse foramen of C1, along the lateral lamina of C1 in the suboccipital triangle (bordered by the superior oblique, inferior oblique, and rectus capitis posterior major muscles), reaching the sulcus arteriosus of C1 and then courses superomedially to pierce the dura medial to the occipital condyle (**Fig. 16.3**). This segment is surrounded by a periosteal sheath which blends into the dura as the vertebral artery transitions from extradural to intradural. Thus, the artery is tightly tethered at the point where it pierces the dura, and this is important to note whenever transposition of the vertebral artery is considered. The intradural V4 segment of the vertebral artery passes superior to the roots of C1, and anterior to the posterior spinal artery, the dentate ligament, and the spinal portion of cranial nerve XI (**Fig. 16.4**). Here, it is important to note that the dentate ligament is often attached to the dural cuff where the vertebral artery pierces the dura.

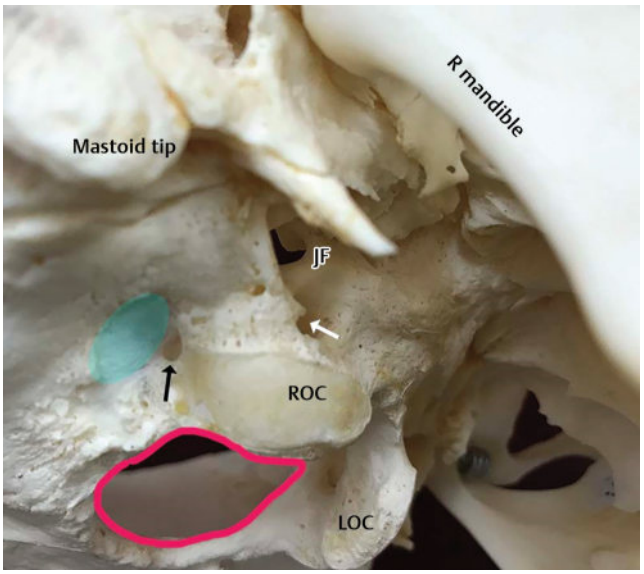


Fig. 16.2 Image of the exocranial surface on the right side. The margins of the foramen magnum have been highlighted in *pink*. *Black arrow*, posterior condylar canal; *white arrow*, outlet of the hypoglossal canal; *blue oval*, right condylar fossa; JF, outlet of the jugular foramen; ROC, right occipital condyle; LOC, left occipital condyle.

Anatomical classification of foramen magnum meningiomas is important, because, even though it does not dictate a specific surgical approach, it influences the overall surgical plan. According to our modification of the original George classification, the lesion in our patient was type A. In this type, the tumor grows caudal to both the vertebral artery and lower cranial nerves, and rarely encases any of these. As opposed to type B tumors above the vertebral artery or type C tumors traversed by the vertebral artery, the position of the lower cranial nerves with type A meningiomas is rarely in doubt, as they are always pushed superiorly and posteriorly.

Options for Approach

The posterior midline approach is frequently used for intradural lesions situated in upper spinal canal, as well as posterior or

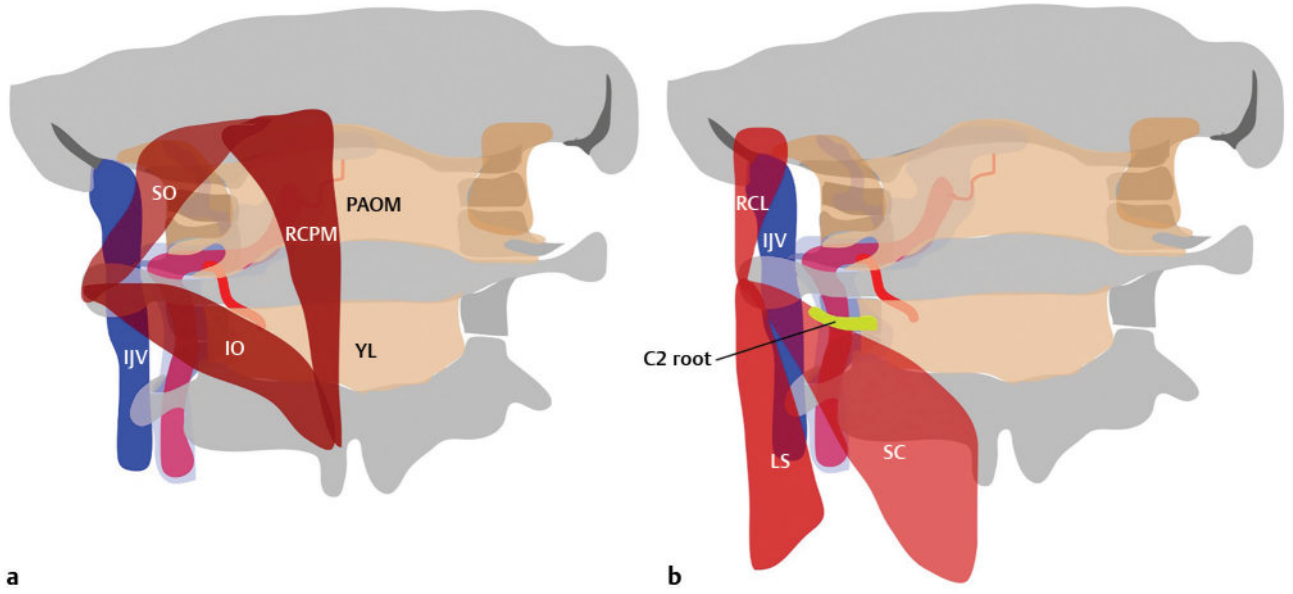


Fig. 16.3 (a) Illustration delineating the relationship between the vertebral artery and the suboccipital triangle. IJV, internal jugular vein; IO, inferior oblique muscle; PAOM, posterior atlanto-occipital membrane; RCPM, rectus capitis posterior major muscle; SO, superior oblique muscle; YL, yellow ligament. **(b)** Illustration demonstrating the deep muscles that attach to the transverse process of C2. IJV, internal jugular vein; LS, levator scapulae muscle; RCL, rectus capitis lateralis muscle; SC, splenius cervicis muscle. (Reproduced from Goel A, Cacciola F. The Craniovertebral Junction. Diagnosis–Pathology–Surgical Techniques. Surgical Anatomy of the Suboccipital and Posterior Paravertebral Muscles. 1st ed. Thieme; 2010.)

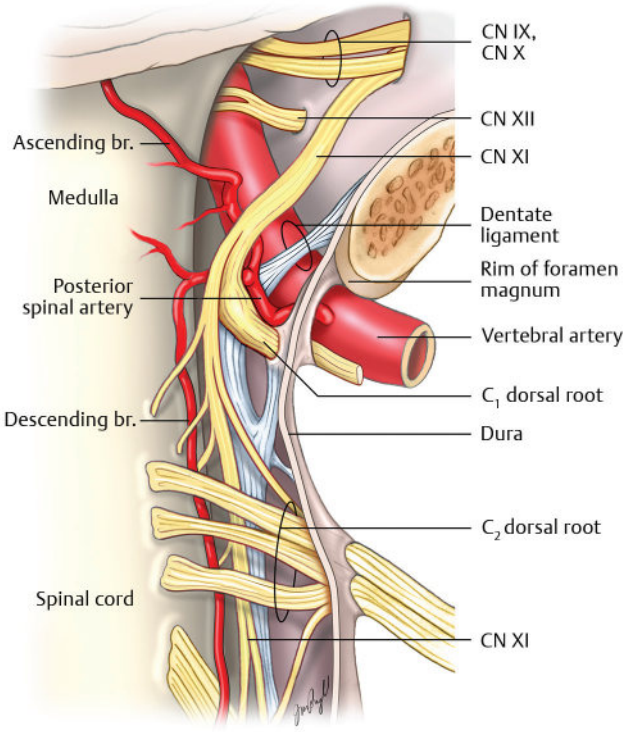


Fig. 16.4 Detailed view of foramen magnum, right side. The spinal portion of the accessory nerve arises from the dorsolateral margin of the spinal cord and ascends between the dorsal roots and the dentate ligament. The most rostral triangular process of the dentate ligament is attached to the dura at the level of the foramen magnum, and is often integrated with the cuff where the vertebral artery pierces the dura. This part of the dentate ligament should be cut to increase exposure to the tumor.

posterolateral areas above the foramen magnum. For ventrally located lesions such as in our patient, the problem with this approach is that the brainstem and spinal cord would obstruct the surgeon's line-of-sight, and accessing the target would therefore require unacceptably forceful retraction on these delicate structures.

Alternatively, anterior approaches, whether by a microscopic transoral or endoscopic endonasal approach (EEA) can be reasonably considered for approaching lesions in front of the brainstem at the foramen magnum. The main advantages of these approaches are direct access to the target obviating any need for retraction, and early elimination of the vascular supply of the tumor as its dura attachment is eliminated before the tumor itself is reached. One major disadvantage of these approaches is that the operation is performed through a contaminated surgical field and the risk of meningitis is therefore quite high. The potential of cerebrospinal fluid (CSF) leakage raises this risk even further. As such, even though resection of clival chordomas using EEA had been frequently reported, resection of meningiomas with wide dural attachments, requiring removal of the anterior parts of the atlas and axis, are rarer in the literature. Specifically, for our patient, the endoscopic endonasal transclival approach may have difficulty reaching the inferior portion of the tumor as it extended nearly to the bottom of C2.

Approach of Choice

For ventrally located tumors at the foramen magnum, our preference is to use the far-lateral approach with varying degrees of condylectomy. In fact, the need for condylectomy is inversely proportional to the tumor size. Although this may run against intuition, a large tumor pushes the medulla and spinal cord significantly backward, and in so doing, creates a space between the occipital condyles and medulla where it is vulnerable to attack (a "weak side"). Cutting the dentate ligament untethers the upper spinal cord and rotates it further out of harm's way. For this reason, we have found that with large type A foramen magnum meningiomas, we can usually spare the condyle.

One potential disadvantage of this technique is that "early" detachment of the tumor from its dural base, which otherwise would retain the tumor less bloody, is not feasible in this approach. However, the tumor can still be devascularized with meticulous hemostasis as the resection proceeds piecemeal, and surgical freedom increases during the operation as the bulk of the mass is stepwise removed.

Questions

1. During the soft tissue dissection above and around the lamina of C1, how does one locate the VA?
2. How does one stop the bleeding from the condylar vein and venous plexus around vertebral artery?
3. How should the lower cranial nerves be handled on the surface of the tumor?

■ Description of the Technique

The patient was placed in the lateral position right side up, with the cervical spine in neutral position without flexion or rotation. The vertex of the head was tilted 30 degrees toward the floor. The right shoulder was retracted caudally by restraint strap to enlarge the space between occipital bone and cervical spine. Intraoperative neurophysiological monitoring was used per routine (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral
Craniotomy: suboccipital
Modifier: retrocondylar

The "hockey stick" skin incision extended from the root of the mastoid process along the line 1 cm below the superior nuchal line to the inion, and downward in the midline to the C3 spinal process of axis. The strict midline plane dissection down to the periosteum of occipital bone and laminar arches of C1 created a bloodless approach. The myofascial flap was retracted dorsolaterally. The muscles were stripped off the posterior arch of C1, from medial to lateral, with care not to damage the vertebral artery in the suboccipital triangle. Moving laterally, when the C1 lamina changes from a relatively flat surface into an edge, one knows that this is indicative of the sulcus arteriosus and the vertebral artery is just above. Blunt dissection using dissectors

wrapped by gauze is our technique of choice to strip the muscle in the subperiosteal plane. As the venous plexus around V3 is enclosed in the periosteal sheath, staying subperiosteal can reduce venous bleeding. If bleeding does occur from the venous plexus or posterior emissary vein, a combination of carefully bipolar coagulation, Gelfoam and bone wax usually controls it readily (**Fig. 16.5**).

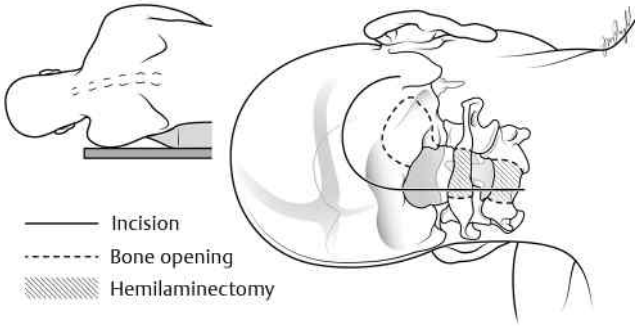
Operative Setup

Position: lateral

Incision: "hockey stick"

Bone Opening: right suboccipital

Durotomy: from C2 to posterior fossa, based laterally



A right-sided suboccipital craniotomy and laminectomy of C1 and C2 were performed in standard fashion. The opening at the foramen magnum was then enlarged up to the posterior edge of the right occipital condyle and the medial edge of the distal sigmoid sinus. The condyle emissary vein and posterior condylar canal were useful landmarks to locate the posteromedial occipital condyle, where the bony removal stopped. The dura mater was opened in a curve pattern, based on sigmoid sinus and posterior margin of condyle, from the C2 to the superolateral apex of the craniotomy (see **Operative Setup**). As soon as the dura was opened, the attachment of the upper triangular processes of the dentate ligament was identified and divided to increase the exposure (**Fig. 16.6a**).

As predicted from the tumor's anatomical classification, the tumor was found anterior to the glossopharyngeal, vagus, and accessory nerves, C1 nerve root, and the lateral medullary segment of the vertebral artery. The vertebral artery was visually traced, and its position was noted for its protection. Debulking the tumor through the interval between the rootlets of C1 and C2, and between C1 and the lower cranial nerves proved difficult as the working space was quite narrow. Dural detachment of the tumor was performed dorsally to ventrally only in areas with enough space for insetting the bipolar forceps (**Fig. 16.6c**). The arachnoid interface between brainstem and the meningioma was intact in most areas, and where identifiable, sharp dissection was used. As piecemeal removal of the tumor progressed, the working space grew to permit working between nerves previously pressed together. Preoperative MRI scans showed the semi-encasement of the right vertebral artery by the lesion, but with identifiable subarachnoid space that suggested no adhesion between the lesion and the vertebral artery. The vertebral artery was predictably displaced superiorly. Both posterior inferior cerebellar arteries, which had been clearly identified in the MRI scans, were free from involvement by the lesion. Carefully, feeding vessels of the tumor were identified as such and coagulated. In this manner, with piecemeal removal and meticulous hemostasis along the way, the tumor was removed completely (**Fig. 16.7**). The dura mater was closed watertight with a fascia lata graft instead of dural substitute, and the rest of the closure was done in standard fashion.

Surgical Pearls

1. Only a minority of foramen magnum meningiomas are strictly anterior, and the majority of them deviate to one side or the other. This makes the far-lateral approach usable in most patients. Meanwhile, most symptomatic patients harbor moderate-to-large tumors, and by displacing the medulla, these tumors "create" the corridor for their very own exit.
2. Condylectomy, by removing more bone, provides additional working space for tumor resection, and mitigates the need for brainstem retraction. It is most suitable for small, anteriorly located tumor, which does not "create" its own wide corridor.
3. Encasement of blood vessels must be evaluated carefully before the operation. If the vertebral artery is encased, separation of one end of the vertebral artery from the tumor should be completed first, then repeated for the other end. From the ends, one works toward the middle of the encasement. In cases in which the tumor invades the adventitia, or severely narrows the lumen, a bypass operation or leaving a cuff of tumor behind attached to the vessel, may be considered.
4. When encountering subpial infiltration of the tumor to the brainstem, separating the interface between tumor and brainstem may be impossible. In particular, forceful separation of adhesions to the ventral part of brainstem can cause motor weakness. In this case, leaving tumor residual may be necessary and sensible.
5. Drilling the C1–C2 facet joint, removing the lateral mass of C1, transposing the vertebral artery out of the transverse foramen of C1, in addition to condylectomy, can all increase exposure ventral to the brainstem. However, all of these maneuvers can add to the morbidity of the operation, and their necessity must be carefully evaluated before and during surgery.

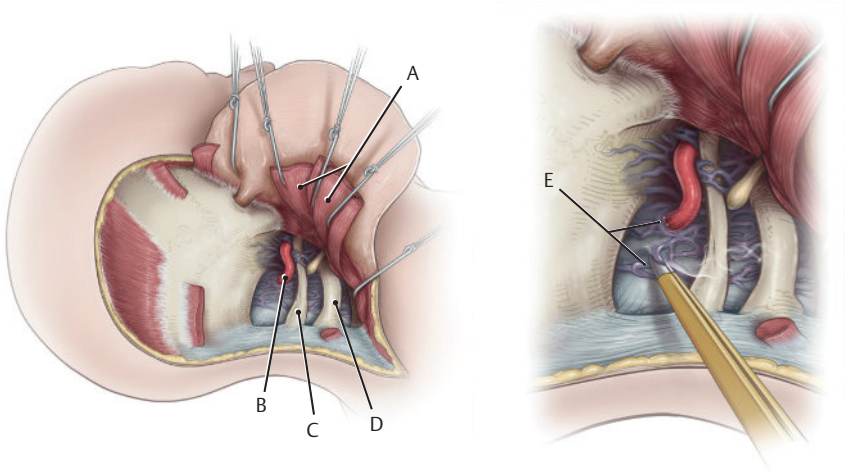


Fig. 16.5 A hockey stick incision was used, starting at the mastoid prominence and proceeding under the superior nuchal line to the midline. The muscle was freed along the nuchal line, leaving a 1-cm edge of nuchal fascia and muscle for closure. The incision was extended caudally down to the midcervical level. The muscle flap was dissected from the suboccipital bone and the C1 and C2 laminae. The muscle flap was reflected inferiorly and laterally with fishhook retractors to expose the lateral mass of C1 and the vertebral artery from C1 to its dural entry. If venous bleeding is encountered, electrocautery is carefully used to coagulate the plexus associated with the vertebral artery to avoid injury to the vertebral artery. A, cervical muscle (reflected); B, vertebral artery; C, C1; D, C2; E, venous plexus.

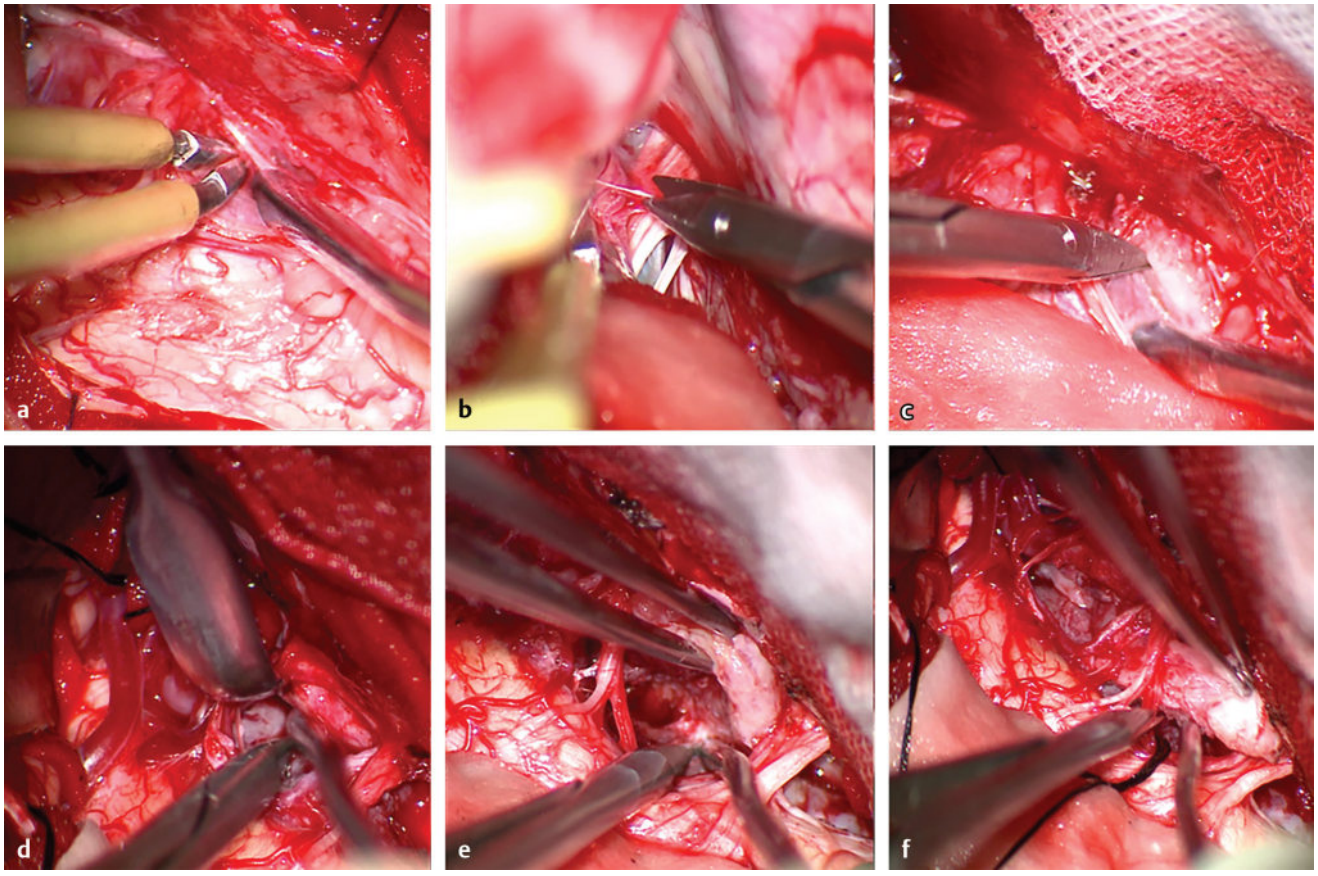


Fig. 16.6 Intraoperative images of the surgical procedure. The tumor was exposed via a far-lateral retrocondylar approach. **(a)** Dissection of the dural attachments by bipolar cautery and sharp dissection. The dentate ligament was cut. **(b)** The arachnoid attachment between the tumor and lower cranial nerves was separated. **(c)** The detachment of the tumor continued in a piecemeal manner and debulking increased the surgical freedom. **(d)** The ipsilateral vertebral artery was elevated to peel the tumor from the adventia of the vessel through the intervals between nerve rootlets. **(e)** The tumor was removed piece by piece with sharp dissection. Forceps were used to remove tumor chunks. **(f)** After completely separating the tumor from surrounding structures, the tumor was removed via the interval between nerve rootlets.

■ Aftercare

The patient suffered palsies of cranial nerves IX to XII after surgery and had mild swallowing dysfunction and tongue deviation to right. As such, for airway protection, the endotracheal tube was kept for 4 days, after which he was extubated without problems. During the postoperative course, dysphagia improved but not completely, and he was fed by nasogastric tube for 9 days before the tube was removed. No other complications were observed. The patient was discharged 14 days after resection. Pathological examination confirmed a diagnosis of a transitional meningioma (WHO grade I), and radiotherapy was not necessary. At recent follow-up, deficit of cranial nerves IX to XII were improved. Postoperative MRIs showed a complete resection without recurrence to date (**Fig. 16.8**).

■ Possible Complications and Associated Management

Surgical morbidity associated with foramen magnum meningiomas is dependent on the location and the radiographic classification of the lesion. Lower cranial nerve deficits are relatively uncommon in type A lesions because of the inherent spatial relationship that had been described by Bruneau and George, as well as Li et al. Aside from lower cranial nerve problems, the most common morbidities include pyramidal signs and CSF leak.

In patients with long-standing symptoms, compensation by the contralateral lower cranial nerves can occur, reducing the impact of postoperative ipsilateral lower cranial nerve palsies. For this reason, patients with recent and progressive symptoms, and elderly patients are most vulnerable to lower cranial nerve deficits after surgery. Patients should be carefully evaluated for airway protection, and it is our routine to use the following criteria: (1) consciousness; (2) tongue extension test; (3) pharyngeal reflex; (4) voluntary coughing; and (5) passive coughing before considering extubation. Tracheostomy should be liberally considered if there is doubt.

Since most lower cranial nerve deficits are transient, gross tumor resection should be considered in most young and healthy patients at initial presentation if it can be accomplished without neurovascular injury. However, in cases where there is brainstem edema, invasion of the brainstem or arterial adventitia, complete encasement or narrowing of a major artery, partial resection should be seriously considered. Preservation of neurological function and quality of life are paramount especially since the tumor is benign and evolves only slowly.

Surgical mortalities are rare but can be due to respiratory failure, vertebral artery injury, brainstem infarction, or aspiration pneumonia. Retraction of the brainstem should be strictly avoided, and perforating arteries to the brainstem should be preserved as carefully as possible. The surgical goal should be tailored to each patient's individual preoperative findings and clinical status, and in the appropriate patients, a subtotal resection may be the most rational goal to preserve the quality of life.

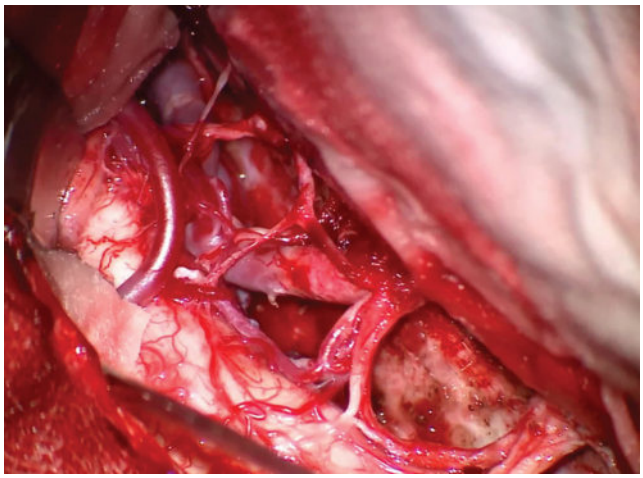


Fig. 16.7 Intraoperative view after complete tumor resection.

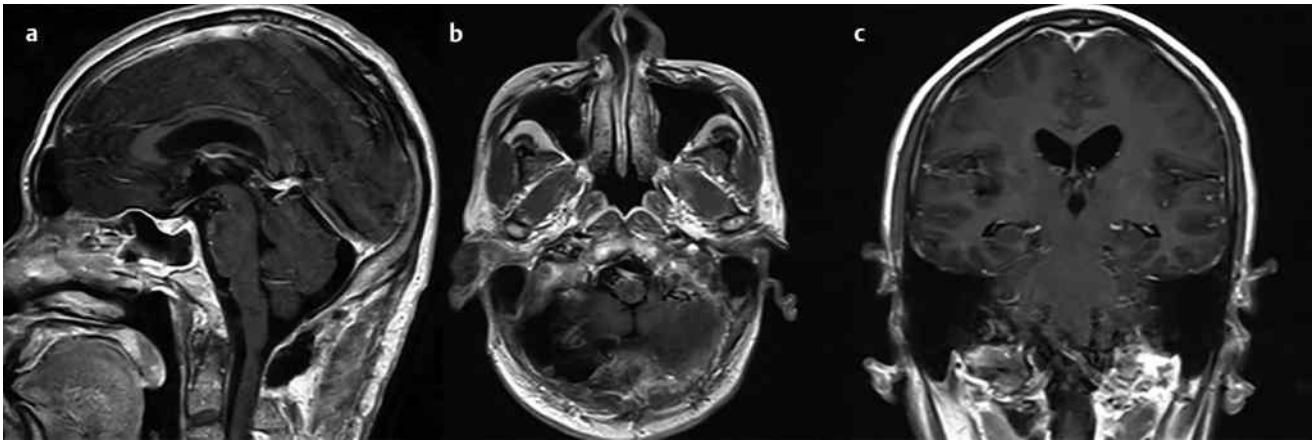


Fig. 16.8 Postoperative MRI. (a–c) The images show a complete tumor resection.

Perspective

Wei-Hsin Wang and Juan Carlos Fernandez-Miranda

■ Introduction

From an anatomical standpoint, foramen magnum meningiomas can be classified in the axial plane as ventral, lateral, and dorsal in relation to the cervicomedullary junction. They can also be classified in the sagittal plane into predominantly spinal or predominantly clival, based on their craniocaudal extension in relation to the foramen magnum. For tumors located along the ventral aspect of the foramen magnum, particularly when they have predominant clival extension, surgical resection is always challenging. As presented previously in this chapter, the lateral extension of the suboccipital approach—the far-lateral approach—is widely used for foramen magnum meningiomas with proven safety and efficacy. However, when utilized for tumors with ventral and predominantly clival extensions, it carries a considerably high risk of transient or permanent dysfunction of the lower cranial nerves.

It is precisely for this reason that the ventral approaches to the foramen magnum, such as transoral and transnasal approaches, were introduced as alternative choices that provide direct access to this region, minimizing the manipulation of the lower cranial nerves, cervicomedullary junction, and vertebral arteries. With increasing experience and technical development, the EEA has become an attractive option for selected foramen magnum meningiomas. Precise anatomical knowledge, meticulous endoscopic technique, and extensive experience with reconstruction of skull base defects are paramount for the successful application of the EEA for these challenging tumors.

■ Case Presentation

A 46-year-old woman presented with significant neck pain as well as numbness in both arms lasting several months. Imaging studies revealed a large tumor along the ventral foramen magnum with predominantly clival extension causing severe compression of the medullary region. The most likely diagnosis was a foramen magnum meningioma (**Fig. 16.9**).

■ Anatomical and Therapeutic Considerations

This tumor was predominantly located at the ventral aspect of the foramen magnum, and its inferior border was at the level of the superior aspect of C1 anterior arch, without any further caudal extension. The vertebral arteries were located laterally but the right vertebral artery was partially encased.

Options for Approach

In general, surgical options to ventral or ventrolateral aspects of the foramen magnum include posterior, posterolateral, and ventral approaches. The suboccipital midline approach was initially used, but the high rates of morbidity and mortality related to brainstem retraction limited its application to dorsal foramen magnum meningiomas. The far-lateral approach, with or without partial condylectomy, provides a lateral-to-medial trajectory and a better route to the ventral region. It is an ideal approach for lateral and ventrolateral meningiomas, and for those with caudal extension beyond C1, because the tumor often opens working space in between neurovascular structures.

The foramen magnum meningioma presented in the previous section extended below C1 (stopping at the level of C2), and created a surgical corridor below the dural entry point of the vertebral artery. It also extended into the ventral foramen magnum by displacing lower cranial nerves superoposteriorly. As such, we agree with the authors that these are conditions quite favorable for a far-lateral approach. However, the real challenge for any posterolateral approach is in handling ventral meningiomas that are above C1, extending superiorly into the lower clival region with posterolateral displacement of the neurovascular structures. In such cases, there is a narrow working corridor, requiring gentle manipulation of the vertebral artery, medulla, and cranial nerves IX to XII. This can be expanded with condylectomy, but only to a limited degree without destabilizing the craniocervical junction.

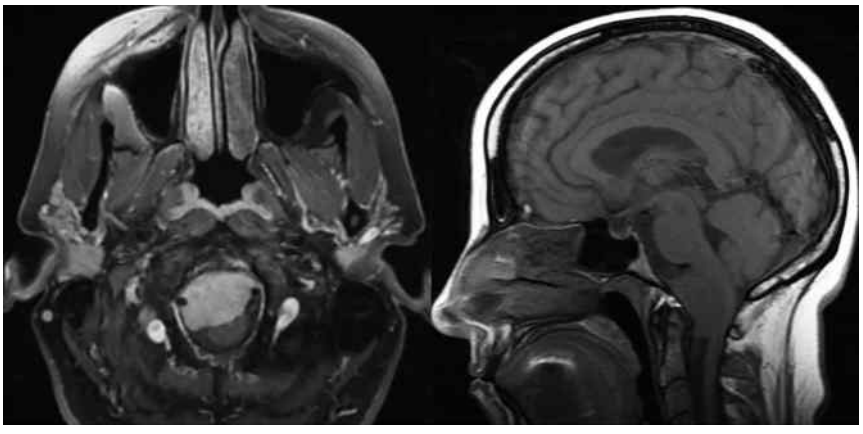


Fig. 16.9 Preoperative MRI revealed tumor located at the ventral aspect of foramen magnum.

Approach of Choice

In our case, we selected the EEA mainly due to its ventral location and predominantly superior/clival extension. When the lesion is mostly ventral to the pons and medulla, in between the vertebral arteries, hypoglossal nerves, and lower cranial nerves, the EEA provides direct access to the lesion via anterior-to-posterior and medial-to-lateral trajectories that may prevent the morbidity related to neurovascular manipulation during a lateral-to-medial approach. Furthermore, the EEA corridor exposes the dural attachment of the tumor first, allowing for early devascularization before tumor debulking (**Fig. 16.10**).

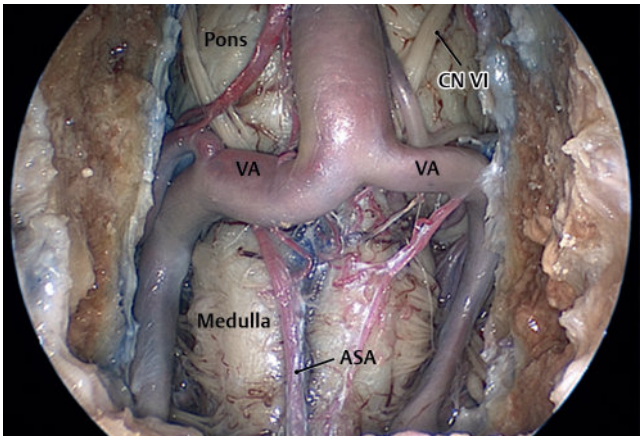


Fig. 16.10 Close-up view of a cadaveric endoscopic transnasal exposure of the lower clival region. This exposure may be further expanded laterally through the jugular tubercle or the occipital condyles or inferiorly through the foramen magnum, according to the pathology. ASA, anterior spinal artery; CN VI, abducens nerve; VA, vertebral artery. (Reproduced from Spetzler R, Kalani M, Nakaji P, eds. Neurovascular Surgery. 2nd ed. Thieme; 2015.)

■ Description of the Technique

The patient's head was fixed in slightly flexed position and rotated toward the surgeon. An extended nasoseptal flap was harvested from the most favorable side without prominent septal spurs. Posterior third septectomy provided binaural access. The exposure extended from the floor of sphenoid sinus superiorly to C1 anterior arch inferiorly. The sphenoidotomy may not be necessary, but the flattening of the maxillary crest to the level of the hard palate is required for more inferior access. The mucosal and muscular layers were elevated and resected together from the inferior clivus by needle cautery and blunt dissection leaving a small cuff inferiorly. When wide access is needed, performing a nasopharyngeal flap or a midline incision is generally avoided because they limit the working space considerably.

The atlanto-occipital membrane was resected to expose the foramen magnum and C1 anterior arch (**Fig. 16.11a**). The upper half of C1 arch and odontoid tip can be drilled without causing instability. The lateral limits of the inferior clival bone drilling were, from superior to inferior: foramen lacerum, petroclival fissure, jugular tubercle, hypoglossal canal, and occipital condyle (**Fig. 16.11b**). In order to expose the lateral wall of the foramen magnum, bilateral medial condylectomies were required. An imaginary line extending inferiorly from the foramen lacerum was used to estimate the lateral limit of the medial condylectomy superficially. While drilling deep, the anterior cortical bone of the intracranial aspect of the hypoglossal canal was encountered and kept intact to prevent injury of the nerve rootlets behind. Less than 20 to 25% of the condyles were resected in this fashion and there was no risk of related craniocervical instability in our experience.

The dural opening was started from the midline and then extended bilaterally. The basilar plexus between the two layers of dura can be easily controlled by hemostatic agents, and

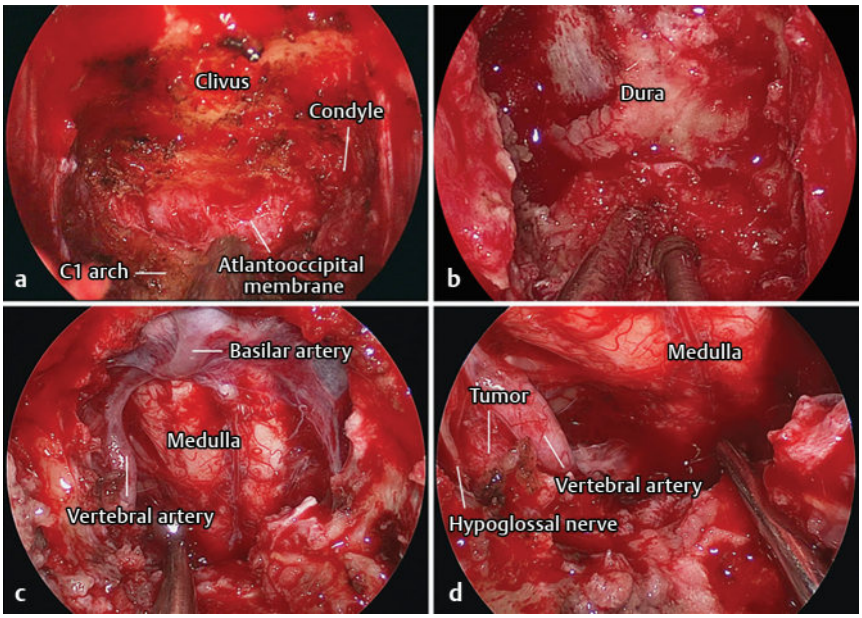


Fig. 16.11 Intraoperative images. **(a)** The mucosal and muscular layers had been resected to expose the inferior clivus and foramen magnum. **(b)** The clival bone and medial occipital condyles had been removed to expose the dura and anterolateral aspect of foramen magnum. **(c)** Nearly total tumor resection was achieved and the neurovascular structures were well preserved. **(d)** A 45-degree angled endoscopic view of foramen magnum showed a small residual tumor with adhesion to the hypoglossal rootlets.

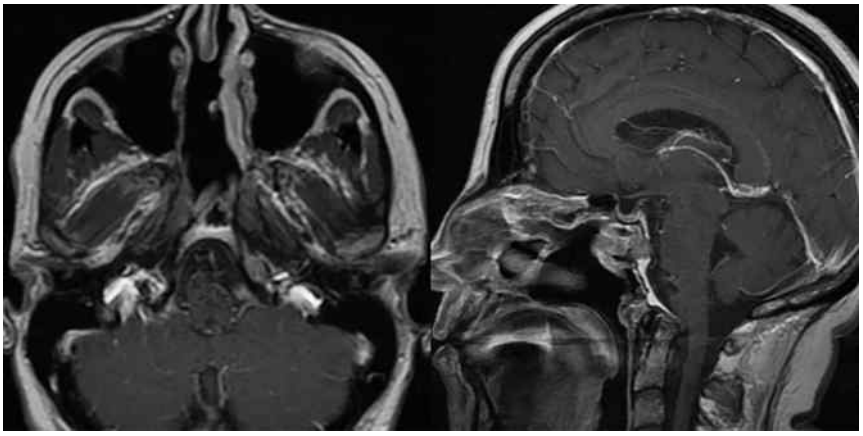


Fig. 16.12 Postoperative MRI. Images after surgery revealed a complete resection and brainstem decompression. The enhancement of the nasoseptal flap in MRI provided the evidence of good flap vascularization and robust reconstruction.

is typically not prominent at this low location. Extensive dural resection is ideal for meningiomas to facilitate better recognition of neurovascular structures and to achieve Simpson grade I resection whenever possible. Microsurgical techniques were used for dissection and resection of intradural lesions. No manipulation of neurovascular structures was required to access and debulk the tumor.

Extracapsular dissection was attempted only after extensive intracapsular debulking. It is not recommended to attempt en bloc resection of tumors because of the risk of damaging surrounding neurovascular structures. The lower cranial nerves were identified superiorly and the hypoglossal nerves laterally, just behind the vertebral arteries on each side. The tumor was then carefully dissected off the right vertebral artery (**Fig. 16.11c**). A very small portion of the tumor extended into the right hypoglossal canal and was adherent to intracanalicular hypoglossal rootlets. This was left as an intended residual to prevent nerve injury (**Fig. 16.11d**).

Reconstruction was completed by a multilayer technique, including inlay collagen layer, onlay fascial lata graft, fat graft reinforcement, and extended nasoseptal flap. A lumbar drain was used to drain 10 cc CSF per hour for 3 days. The use of a lumbar drain is recommended because it has shown to significantly reduce the incidence of postoperative CSF leaks for transclival defects.

■ Aftercare

No CSF leak occurred in this patient. She was discharged on postoperative day 5 without any neurological deficits. Preoperative upper extremities' numbness completely improved. One year after surgery, she underwent Gamma Knife radiosurgery for the small stable residual tumor. At 5 years' follow-up, she remains asymptomatic without evidence of tumor growth or craniocervical instability (**Fig. 16.12**).

■ Commentary

Surgical resection of ventral foramen magnum meningiomas is challenging due to their deep location and surrounding complex neurovascular structures. Compared to the transcranial far-lateral approach, the main advantage of the EEA is its direct access and less neurovascular manipulation. For deep intradural lesions, adequate exposure is required for safe resection of tumors. In our previous studies, the inferior transclival approach with medial transcondylar extension provides excellent access to the lateral wall of the foramen magnum. However, lesions that extend below the arch of C1 are contraindicated for EEA because they would require resection of the odontoid body and transverse ligament. Similar to an extensive condylectomy, this would make the craniocervical junction unstable.

The main disadvantage of the EEA is the higher rate of postoperative CSF leakage. The high flow of CSF in the premedullary cistern and the deep location of the lower clivus and the foramen magnum make reconstructions very challenging. Although multilayer techniques plus lumbar drain have significantly improved the rate of postoperative CSF leakage, the techniques for reconstructions of low clival and foramen magnum defects are still evolving and require continued improvement. Patients may not always present with rhinorrhea but with a constant postnasal drip or an increase of intracranial pneumocephalus instead. Early diagnosis and treatment of CSF leaks with return to the OR for further reconstruction is the best way to prevent related meningitis and other potential complications. When needed, a lateral nasal wall/inferior turbinate flap can be used as a rescue flap to reinforce a failed reconstruction.

In conclusion, the EEA provides a privileged route to resect ventral midline skull base lesions including properly selected foramen magnum meningiomas, and may decrease the risk of neurovascular complications. However, the learning curve of the EEA is steep and the potential for reconstruction-related complications is higher than with transcranial far-lateral approaches.

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17 Craniovertebral Junction

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Keywords: craniovertebral junction, chordoma, transcondylar approach, endoscopic endonasal approach, transpterygoid extension

■ Case Presentation

A 33-year-old man with no previous medial history presented with dysarthria, dysphonia, and right hypoglossal palsy. On endoscopic examination, right vocal cord palsy was documented. A head computed tomography (CT) scan was ordered and a large lytic mass located in the lower clivus and right occipital was found (**Fig. 17.1**). A subsequent magnetic resonance imaging (MRI) was done to better delineate all the extensions of the craniovertebral junction (CVJ) mass, and it was found to have a significant intradural extension toward the cerebellomedullary cistern (**Fig. 17.1c, d**).

Questions

1. What is your differential diagnosis based on the MRI and CT appearances?
2. What are the different management options? Is there a role for biopsy?
3. If surgery was considered, what would be your surgical goal and strategy?

■ Diagnosis and Assessment

The imaging characteristics of the lesion, including its location in the lower clivus and CVJ, the destructive invasion of the bone, the irregular calcifications within the tumor, the heterogeneous enhancement in a "honeycomb" pattern, and the high signal intensity on T2-weighted MRI are all highly suggestive of a chordoma. The differential diagnosis includes chondrosarcoma, *ecchordosis physaliphora*, metastatic lesion, and other rare bony tumors (lymphoma, plasmacytoma, etc.).

Considering this patient's symptoms and young age, the high level of suspicion of a chordoma, and the lesion's relatively large size, surgical resection was the treatment of choice. A biopsy might be counterproductive, as it may lead to seeding in the biopsy track. There is ample evidence in the current literature, which shows that gross total resection (GTR) of skull base chordomas improves overall survival and progression-free survival. Furthermore, the first operation is the most important one. In fact, in most surgical series, GTR appears harder to achieve in residual and recurrent disease than during the first surgical attempt. Every effort should thus be made to maximize resection during the first surgery and therefore, GTR was set for the surgical goal in this young patient.

Radiation therapy is not a good first-line option for chordomas. These tumors, though considered to be "benign" histologically, are highly radioresistant, and it is only with very high doses of radiation, often with charged particles, that a

Fig. 17.1 Preoperative images of a CVJ tumor. (a, b) Axial CT images demonstrating osteolysis and mass effect centered on the right C1 lateral mass, right occipital condyle, and lower clivus. **(c)** Axial and **(d)** sagittal T2-weighted MRI demonstrating the tumor extension in the lower clivus, right occipital condyle, and intradurally. There was associated edema in the lower lateral medulla and right cerebellar tonsil (*white arrows*).

biological effect on chordomas can be obtained. Additionally, the target volume is a strong predictor of response to radiation therapy in chordoma, and this patient's tumor was too big for radiotherapy to be a viable stand-alone treatment option.

Preoperative Assessments

Although chordomas tend to be midline lesions and affect preferentially the middle section of the clivus, they also have the tendency to extend locally and invade different compartments and anatomical structures around the skull base. The tumors often have asymmetric growth patterns with multiple loculations. On occasion, this means that the straighter surgical corridor may not be the one that allows the most complete resection. Therefore, before choosing the approach that offers the best chance at the optimal surgical result, one must carefully delineate all the tumor extensions, large or small, and identify them as surgical targets.

Neurological deficits, especially those involving cranial nerves and those that are "fixed," must be considered from a functional point of view, in order to avoid bilateral deficits or those uncorrectable through rehabilitation. Endoscopic assessment of the vocal cords and testing of swallowing function should be liberally considered, and patients should be counseled on the need for tracheostomy and feeding tubes in the postoperative period.

Preoperative MRI should include volumetric *constructive interference in steady state* (CISS), *fast imaging employing steady-state acquisition* (FIESTA), or similar sequences to assess cranial nerves and their relative position and relationship with the chordoma. It can also be useful to determine if there is intradural invasion and to assess the relationship with the vessels of the posterior circulation and the brainstem. A CT is helpful in the assessment of the bony anatomy, including intratumoral calcifications, extent of condylar invasion and destruction, position of the jugular bulb, and possible misalignment of the CVJ. The surgical plan must also take into account factors such as the vascular anatomy and cerebral hemodynamics. A CT angiogram is usually sufficient, unless involvement and/or encasement of a major vessel is seen. When considering exposure and mobilization of the vertebral artery, it is important to exclude the presence of an extradural origin of the posterior inferior cerebellar artery. A cerebral angiogram with a balloon test occlusion of the involved vertebral or internal carotid artery (ICA) may also need to be considered.

■ Anatomical and Therapeutic Considerations

Options for Approach

The main surgical approaches to clival chordomas may be categorized into (1) midline endoscopic and (2) posterolateral "open." The case for the endonasal endoscopic approach (EEA) for chordomas has been made very strongly in the recent literature and the rationale for choosing this approach is convincing. The soft and "suckable" consistency of chordomas, together with their origin in the midline bony structures, make them uniquely suitable as targets for the endoscopic midline approach. One element that often determines if the midline approach is suitable for a specific lesion is the mucosal and soft tissue flaps available for reconstruction. For recurrent tumors, the vascularized mucosal flaps generally used as first-line reconstructive barriers may be used-up or compromised. Since prolonged cerebrospinal fluid (CSF) leak and meningitis are unacceptable complications, the midline approach can only be considered if a vascularized reconstructive barrier remains a viable option.

However, the main limitation of the EEA is the lateral extension of the tumor beyond the level of cranial nerves foramina, including the hypoglossal canal, the jugular foramen, and the internal auditory meatus. In some cases, middle or inferior turbinectomy, maxillary antrostomy, transpterygoid approach, eustachian tube resection, and pharyngeal muscle dissection can extend an endoscopic midline approach (EMA) laterally and inferiorly, but despite these modifications, the reach may still be insufficient. Moreover, these maneuvers can significantly impact sinonasal function and postoperative quality of life. Before choosing one of these modifiers, one must assess the possibility of intradural invasion and adhesion to major blood vessels or the brainstem. The control and management, through the endoscope, of any potential carotid injury must be considered as well.

Posterolateral approaches (PLAs) still have an important role to play in the management of CVJ chordomas. In fact, in recurrent cases, for tumors with significant lateral or intradural extension, PLA may still be the better and safer option. Depending on the height, exact location, and morphology of the lesion, (1) an anterior transpetrosal, (2) a retrosigmoid and/or posterior petrosal, (3) a far-lateral retrocondylar/transcondylar, or (4) an anterolateral approach (ALA) may be chosen. The anterior transpetrosal is favored in lesions located higher up in the clivus and at the petrous apex (usually above the level of the internal acoustic meatus). The retrosigmoid approach is most useful in lesions centered on the cerebellopontine angle. The far-lateral approach allows great posterior exposure of the jugular foramen, as well as of the vertebral artery, the occipital condyle, the lower cranial nerves, and the cerebellomedullary cistern intradurally (**Fig. 17.2a**). The ALA, described by Bernard George in 1988, provides access of extradural and mixed intra- and extradural lesions of the anterior aspect of the lower clivus and C1–C2, with additional access to the vertebral artery and the lower cranial nerves in the neck (**Fig. 17.2b**).

Approach of Choice

In the present case, there are several aspects that made EEA difficult or unsuitable. There was significant intradural extension, as well as cerebellar edema, which was interpreted as the possibility of pial invasion. Since a large dural defect was expected, the increased risk of postoperative CSF leak made EEA less than ideal. There was tumor extension lateral to the level of the jugular foramen and hypoglossal canal, which would be difficult to reach with EEA, and given the destruction of the right occipital condyle, postoperative instability can be expected. All this made the PLA through a "hockey stick" incision the approach of choice, especially since that incision can be reused for CVJ stabilization at a later date.

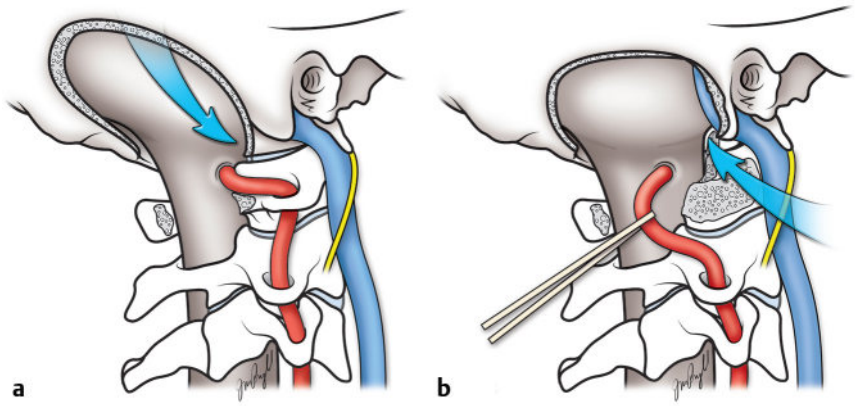


Fig. 17.2 Illustration comparing the posterolateral and anterolateral approach. **(a)** Approach through the posterolateral corridor is depicted here as the far-lateral transcondylar approach. Note that in this depiction, the vertebral artery has been left in the foramen transversarium, but it can be mobilized posteriorly to give more exposure to the occipital condyle. The far-lateral approach offers great exposure and control of the jugular foramen, the vertebral artery, the occipital condyle, the lower cranial nerves, and the cerebellomedullary cistern. **(b)** The anterolateral approach (ALA), described by Bernard George in 1988, provides access to extradural and mixed intra- and extradural lesions of the anterior aspect of the lower clivus and C1–C2. It also grants access to the vertebral artery and the lower cranial nerves in the neck.

However, it may be useful to consider endoscopic and microscopic "open" techniques not as competing, but as complementary, modalities. Indeed, recently, the use of endoscopic assistance has emerged as a very useful adjunct in extending the reach of traditional skull base approaches and improving resections. The advantages of the endoscope are not only improved illumination and visualization in deep locations, but also better appreciation of surgical blind spots.

Questions

1. What anatomical landmarks can be used to assess the risk of CVJ instability postoperatively?
2. What is the venous structure that is encountered before the hypoglossal canal during condylar drilling?
3. Can you name one vascular anatomy variant that is relevant to the dissection done for a far-lateral approach?

■ Description of the Technique

The patient was positioned in the three-quarter prone position with the right side up and head fixed in pins. The head was then further rotated (10 degrees to the left side), flexed and with a slight lateral tilt to improve exposure of the retrocondylar fossa. A slight reverse Trendelenburg positioning of the surgical table was used to ensure good venous outflow and reduce venous bleeding. The abdomen was prepared for harvesting a fat graft to repair the dural defect and fill the void left after drilling of the occipital condyle (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: postero (infero) lateral
Craniotomy: suboccipital, retrosigmoid
Modifier: condyle resection

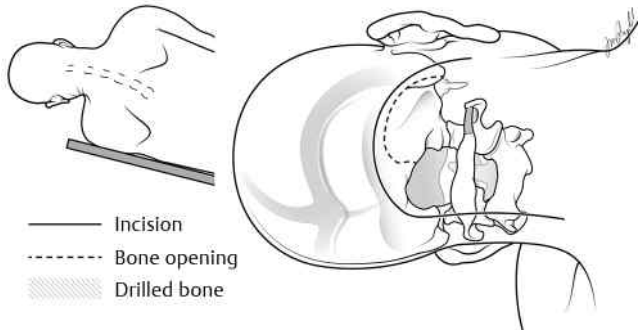
In all our skull base cases, we use surgical navigation with both MRI (T2 weighted or CISS/FIESTA and T1 weighted with gadolinium) and CT imaging fusion. Monitoring of motor evoked potentials and electromyographic monitoring of cranial nerves VII (orbicularis oculi and oris muscles), IX (pharyngeal elevator muscle), X (vocal cord), and XI (trapezius muscle) and XII (tongue) were used.

Opening

A "hockey stick–shaped" incision was made, starting from the tip of the mastoid, going superiorly to the level of the insertion of the nuchal muscles and transverse sinus, tracing back to the midline and then down to the level of the spinous process of C2. The incision must extend low enough to allow adequate mobilization of the musculocutaneous flap in an inferior and lateral direction, a maneuver that enlarges the working space and provides an adequate line-of-sight. This was followed by dissection of the avascular midline plane, exposing the occipital bone and lamina of C1 and C2. The right nuchal muscles were then elevated en bloc in the subperiosteal plane. A muscle cuff was left at its tendinous insertion to facilitate closure (see **Operative Setup**).

Operative Setup

Position: three-quarter prone, head rotated left and tilted down
Incision: "hockey stick," mastoid to C2
Bone Opening: suboccipital to foramen magnum
Durotomy: curvilinear around condyle



A “teardrop-shaped” craniotomy was then performed, above and medial to the condylar fossa and connecting to the foramen magnum (**Fig. 17.3a**). Two burr holes were done on each side of the external occipital crest and another one laterally, behind the sigmoid sinus. These burr holes were then connected with the craniotome and the foramen magnum opened.

A partial mastoidectomy and infralabyrinthine drilling were performed in order to skeletonize the inferior part of the sigmoid sinus and jugular bulb. The tumor capsule was visualized immediately posterior to and below the jugular foramen.

Vertebral Artery Transposition

After identification of the horizontal segment of the vertebral artery along the sulcus arteriosus, the transverse process of C1 was removed and the transverse foramen of C1 was skeletonized with a diamond burr and opened. The V3 segment was then displaced posteriorly and out of the transverse foramen of C1, freeing a corridor to the lateral mass of C1 and the atlanto-occipital joint. The periosteal sheath and vertebral plexus around the vertebral artery were kept intact to protect the vertebral artery, reduce venous bleeding, and minimize the risk of dissection.

Condylar Resection

The retrocondylar fossa was exposed and the posterior condylar emissary vein coagulated and cut. The tumor was readily exposed and was found to have eroded the right occipital condyle almost completely. The condylectomy was completed with a diamond burr. The anterior condylar emissary vein was seen, just posterior to the hypoglossal nerve in the hypoglossal canal, which was exposed shortly afterward superior to the tumor (**Fig. 17.3a**). Drilling of the jugular tubercle was then completed to access tumor located higher up in the midclivus.

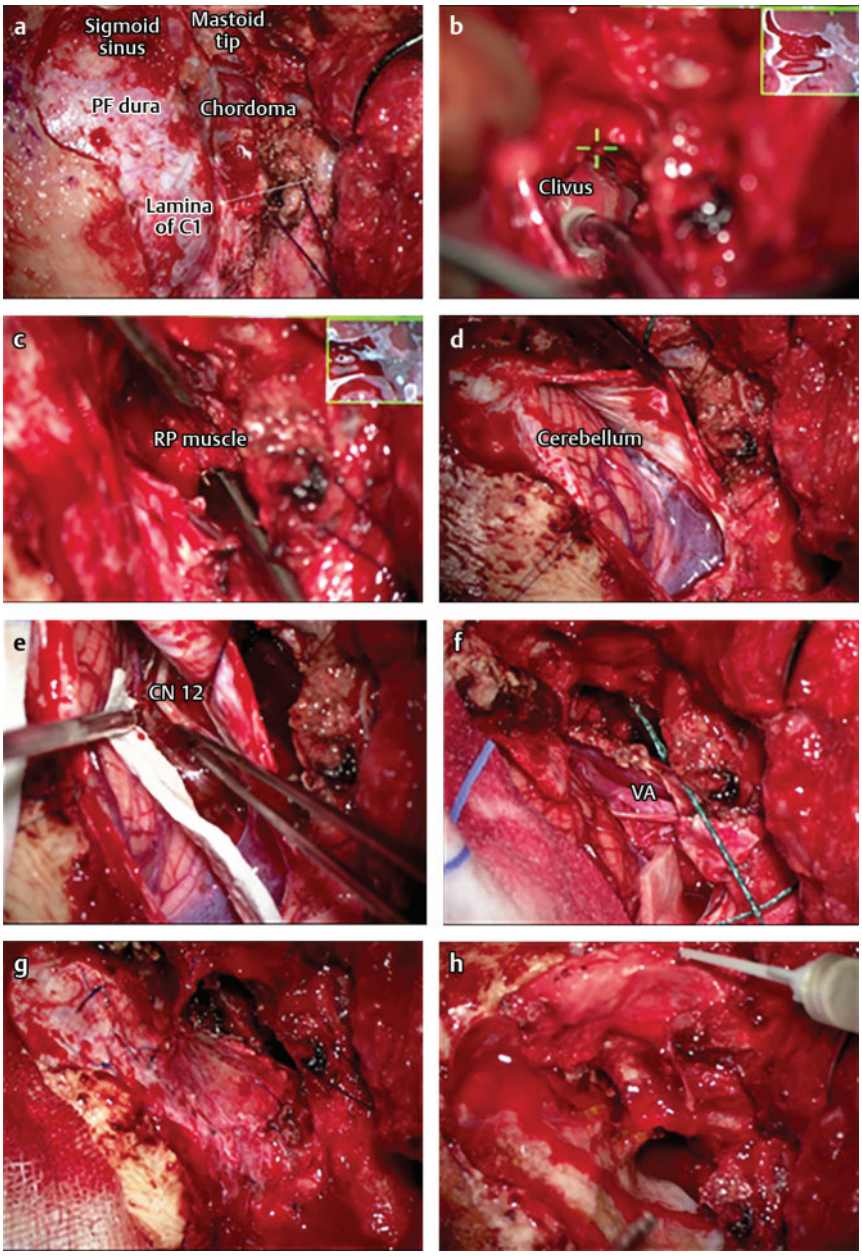


Fig. 17.3 Intraoperative images. (a) Dural exposure after a “far-lateral” craniotomy and skeletonization of the posterior aspect of the sigmoid sinus were performed. The tumor was easily identified and its capsule was preserved during the initial stages of the extradural work. **(b)** After debulking the main tumor mass, the infiltrated clivus was resected with a diamond drill through a window above the hypoglossal canal and below the jugular foramen. **(c)** The lower aspect of the sphenoid sinus was reached superiorly and the retropharyngeal muscles anteriorly, as shown in the neuronavigation inserts at the upper right corner. **(d)** A curvilinear dural incision based on the retrocondylar fossa was used. **(e)** The intradural extension of the chordoma was dissected from the hypoglossal nerve and resected. **(f)** Microscopic resection of the tumor and the invaded dura mater was obtained, including a portion of tumor invading the pia mater of the right cerebellar tonsil. **(g)** Watertight closure of the dura with pericranial graft. **(h)** The mastoid cells were closed with bone wax and covered with fascia and fibrin glue. PF, posterior fossa; RP, retropharyngeal; VA, vertebral artery.

Extradural Tumor Resection

The extradural compartment of the chordomas was thus widely exposed. For the resection, two surgical windows were used: (1) superiorly to the cranial nerve XII and inferomedially to the jugular bulb and (2) inferiorly to the cranial nerve XII and superiorly to the third segment of the vertebral artery. Endoscopic assistance proved useful to see in these deep corridors and remove the chordoma medially to the cranial nerves. After removing all of the tumor in the extradural compartment in the lower half of the clivus, the submuscular retropharyngeal space was exposed (underneath the rectus capitis anterior) (**Fig. 17.3c**), as well as the mucosa of the clival recess of the sphenoid sinus (**Fig. 17.3b**). Looking up with the 30- and 45-degree endoscope, the tumor extending into the petrous apex was resected, and the petrous and paraclival segment of the ICA was visualized from below.

Dural Incision

Next, the posterior fossa and upper cervical dura were incised. This incision was done in a curvilinear fashion and centered on the occipital condyle (**Fig. 17.3d**). Care was taken when crossing the marginal sinus at the level of the foramen magnum as there can be profuse venous bleeding during this step. The tumor was identified, extensively debulked, and dissected from the vertebral artery and small perforators to the brainstem (**Fig. 17.3e**). It was found to be highly adherent to the cerebellum, from which it was dissected in a subpial plane. Fortunately, a clear dissection plane was found between the tumor and the brainstem and lower cranial nerves. GTR of the tumor and wide resection of the involved dura were accomplished (**Fig. 17.3f**).

Closure

Duraplasty with a pericranial graft was supplemented by a fibrin sealant patch (*TachoSil*) (**Fig. 17.3g**). The empty space created by the drilling in the condyle and the preexisting tumor in the clivus were filled with autologous fat and fibrin glue. The opening of the posterior mastoid cells was closed with a thin layer of bone wax, which was then supplemented by a fascia patch and fibrin glue (**Fig. 17.3h**). The bone flap was put back into position and fixed with titanium miniplates and screws. The muscular flap was then attached to the cuff left during the exposure. The skin was closed with staples.

Surgical Pearls

1. The keyhole concept, which has gained interest in cranial neurosurgery, can also be applied to PLAs to the clivus and craniocervical junction. In these areas, because of the muscular and vascular anatomy, small incisions may not be feasible. It is important, however, not to confuse the length of the incision with the concept of minimally invasive surgery. For instance, through a classic “hockey stick” incision and exposure of the occipital bone and condyle, a deep keyhole, located in the condylar fossa for example, can provide access to a large segment of the middle and lower clivus and C1 and C2 vertebrae (**Fig. 17.4a**) with the assistance of the endoscope to navigate this area.
2. The endoscope should be used liberally as a tool to operate in the depths of the triangles defined by the lower cranial nerves. In chordoma cases, this is safely accomplished in the extradural compartment, in the space left by the drilling or tumor-related osteolysis of the condyle and jugular tubercle. Above the hypoglossal nerve, this refers to the space inferomedial to the jugular bulb, and below the nerve, the space above the vertebral artery (V3 segment) (**Fig. 17.4b**).
3. During tumor resection, every caution should be taken to limit tumor seeding in the CSF but even more so through the surgical corridor (noted in as much as 5% of cases). To limit CSF spread, as a general rule, extradural debulking must be maximized before intradural exploration.

■ Aftercare

We do not routinely leave lumbar drains or use antibiotics after surgery, but occasionally have to perform lumbar punctures if patients complain of headaches or if ventriculomegaly is noted on imaging. All patients should have an MRI within 48 hours after surgery to assess the extent of resection and any evidence of complications. The MRI in our patient showed complete resection of the chordoma, and no evidence of ischemia or injury to the brainstem. There was a small area of T2 hyperintensity at the level of the cerebellar amygdala, where the chordoma was dissected in the subpial plane (**Fig. 17.5**). The patient did not suffer any cranial nerve dysfunction or CSF leak. However, he had a postoperative radial neuropathy that was due to positional compression during surgery. This resolved completely within weeks.

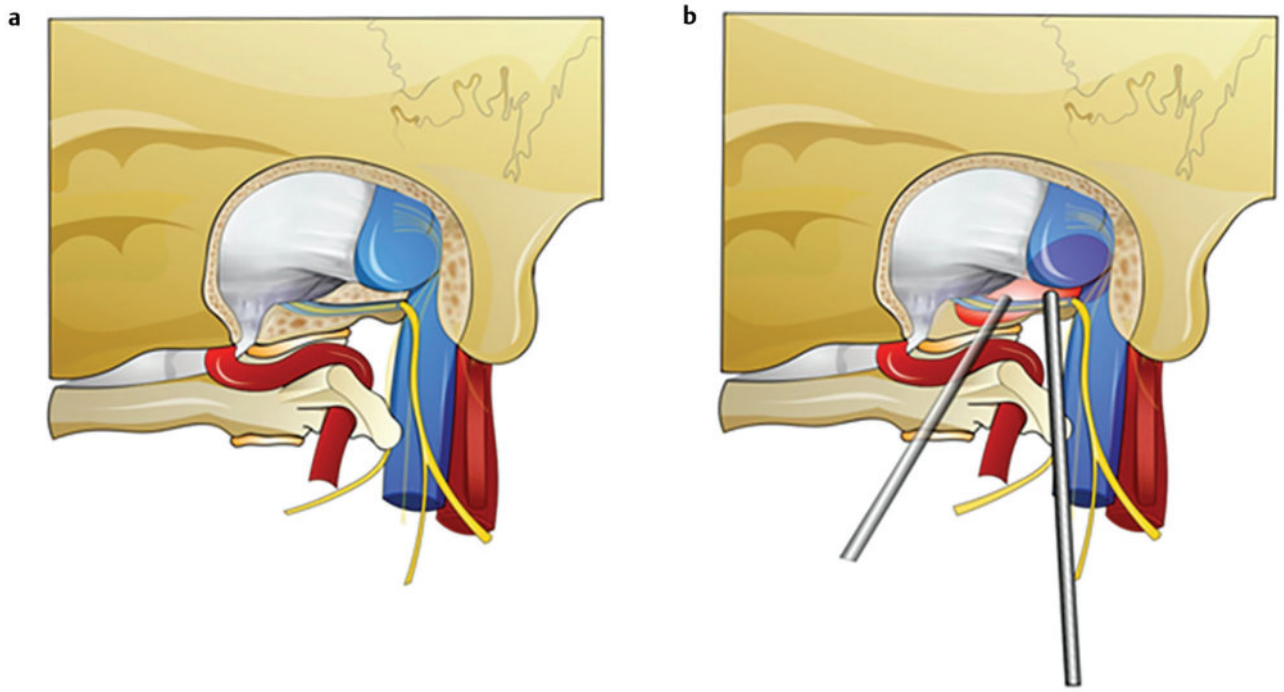


Fig. 17.4 Keyhole concept in the far-lateral approach. (a) In the extradural compartment, two windows can be used to access lesions involving the lower and middle thirds of the clivus and the dens and lateral masses of C1. The first space is located superiorly to the nerve XII, inferomedially to the jugular bulb, and is limited medially by the dura. The second window is inferior to the nerve XII, superior to the third segment of the vertebral artery, and is also bounded medially by the dura. **(b)** Endoscopic assistance can also be used through both windows to improve illumination and visualization in the depths of these surgical windows and "look around corners." In the case of this patient, the vertebral artery was mobilized posteriorly to increase the exposure in the lower window.

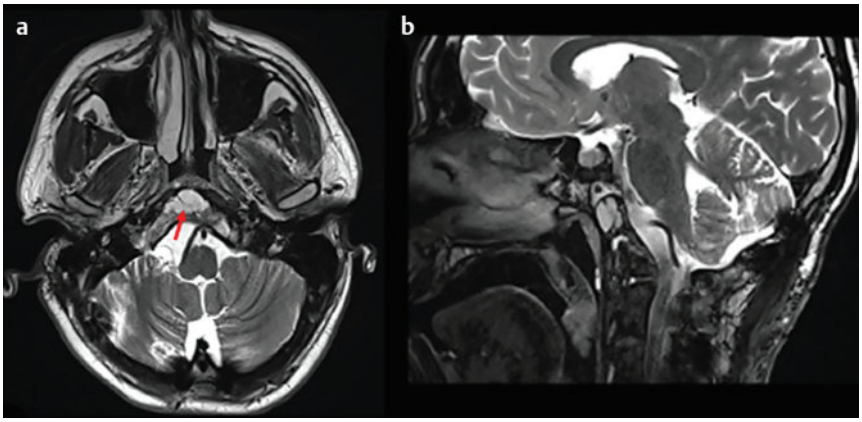


Fig. 17.5 Postoperative MRI. (a) Axial and **(b)** sagittal T2-weighted postoperative MRI demonstrating complete resection. Note the fat graft located in the medial clivus (*red arrow*).

Adjuvant proton beam therapy was recommended. Before and during the radiation therapy, a rigid cervical collar was prescribed because of the extensive condylar resection. After completion of the radiation therapy, a CT scan of the CVJ revealed left-sided head tilt and sagging of the left lateral skull base onto the C1 lateral mass associated with persistent cervical pain (**Fig. 17.6a**). Therefore, an occipital–C5 fusion was thus performed (**Fig. 17.6b**). The strategy of postponing CVJ fusion until after radiotherapy allowed unencumbered proton beam treatment planning.

■ Possible Complications and Associated Management

Postoperative cranial nerve XII palsies are not infrequent, and cranial nerve IX/X palsy may require tracheostomy and/or gastrostomy, hopefully only for the short term. CSF leaks and pseudomeningocele are also frequent after resection of CVJ chordoma. If a CSF leak, our first line of treatment is usually bed rest and lumbar drainage, unless the leak is deemed to be

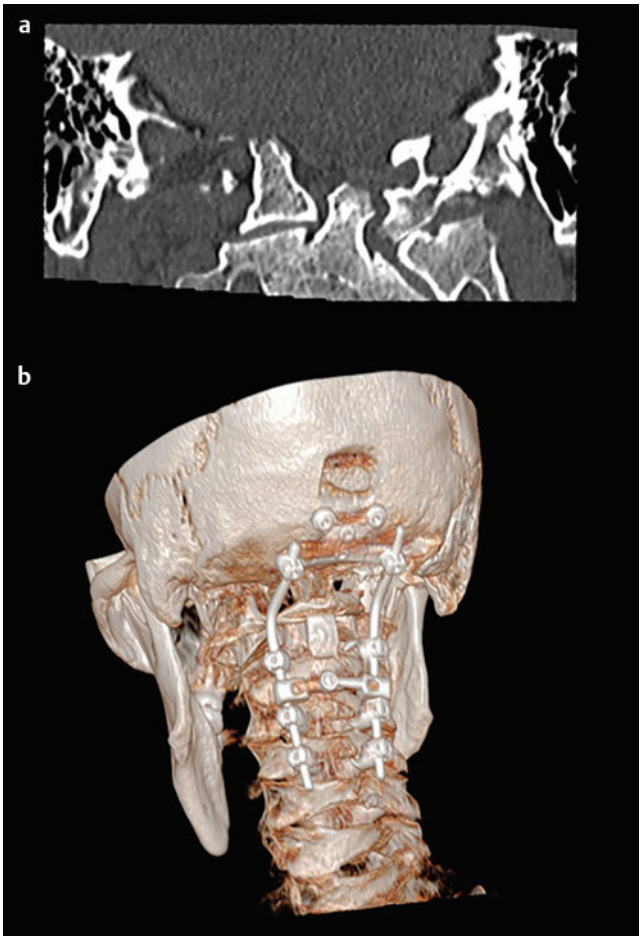


Fig. 17.6 Craniovertebral junction instability and treatment. **(a)** CT scan of the CVJ showing a left-sided head tilt and sagging of the left lateral skull base onto the C1 lateral mass. **(b)** An occipital–C5 fusion was done using sublaminar hooks from C2 to C5.

"high flow," in which case we would reexplore the surgical site. Pseudomeningoceles without leaks often require no further intervention.

In many cases, instrumented fusion is necessary to stabilize the craniocervical junction. If small remnants of tumor were noted on the postoperative MRI, we would on occasion take advantage of the fusion procedure to complete the tumor removal. For CVJ chordomas, it is important to consider the adjuvant treatment plan when designing the occipitocervical fusion and selecting instrumentation, as some implants may hinder proton therapy. We avoid using bulky implants at C1 and C2 such as lateral mass or pedicle screws. In some cases, where instability is occult, such as in the patient described above, it may be beneficial to postpone fusion until after the completion of proton therapy.

Perspective

João Paulo Almeida, Miguel Marigil-Sanchez, Claire Karekezi, and Fred Gentili

Introduction

Chordomas are rare, invasive tumors of the bony and cartilaginous skull base that originate from remnants of the embryonic notochord. Most intracranial chordomas arise from the clivus region, involving the occipital and sphenoid bones, with variable lateral extension into the petrous part of the temporal bone, jugular foramen, and carotid canal. Although generally considered low-grade tumors, they have varying biological behavior, and their central location and tendency to infiltrate bone and encase surrounding neurovascular structures make them surgically challenging. Indeed, attempts to achieve aggressive resection of some invasive chordomas are associated with significant postoperative morbidity.

Contemporary paradigm for managing clivus chordomas is based on maximal safe surgical resection followed by high-dose radiation therapy, usually delivered by proton beam or intensity-modulated radiation therapy (IMRT) to avoid toxic doses to the brainstem. With the development of endoscopic skull base approaches, a less invasive approach option now exists, which allows wide visualization of the clivus region and relatively safe tumor resection (**Fig. 17.7**). Furthermore, by using the more direct midline corridor, the EEA avoids the need of surgical dissection between cranial nerves and mobilization of major intracranial vessels, since these structures are mostly behind the tumor in the anterior-to-posterior surgical trajectory (**Fig. 17.8**). Although ideally suitable for midline extradural chordomas, it is also useful in the management of skull base

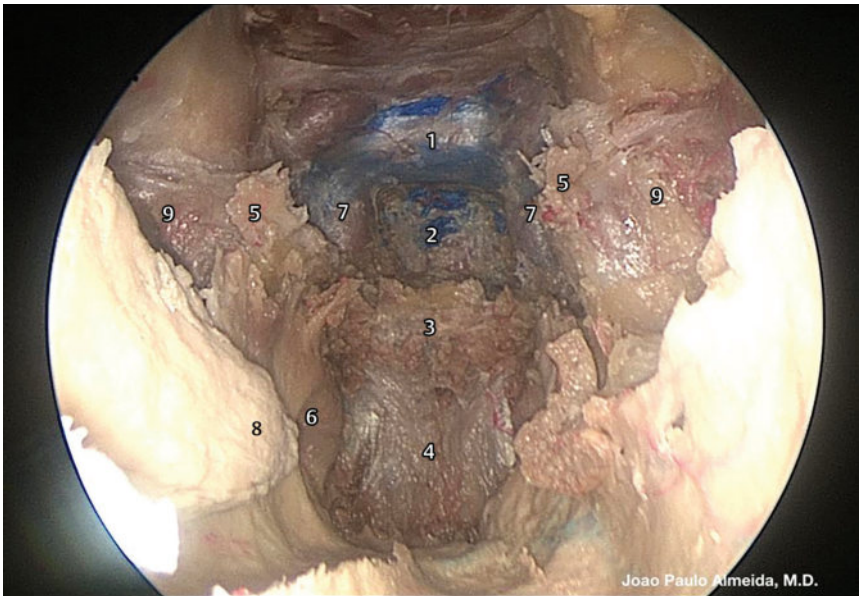


Fig. 17.7 Exposure of the clivus via endoscopic endonasal approaches. 1, sella; 2, clivus recess; 3, floor of the sphenoid sinus; 4, nasopharynx; 5, pterygopalatine ganglion and vidian nerve; 6, eustachian tube; 7, paraclival carotid; 8, inferior turbinate; 9. posterior wall of the maxillary sinus. (Copyright João Paulo Almeida, MD.)

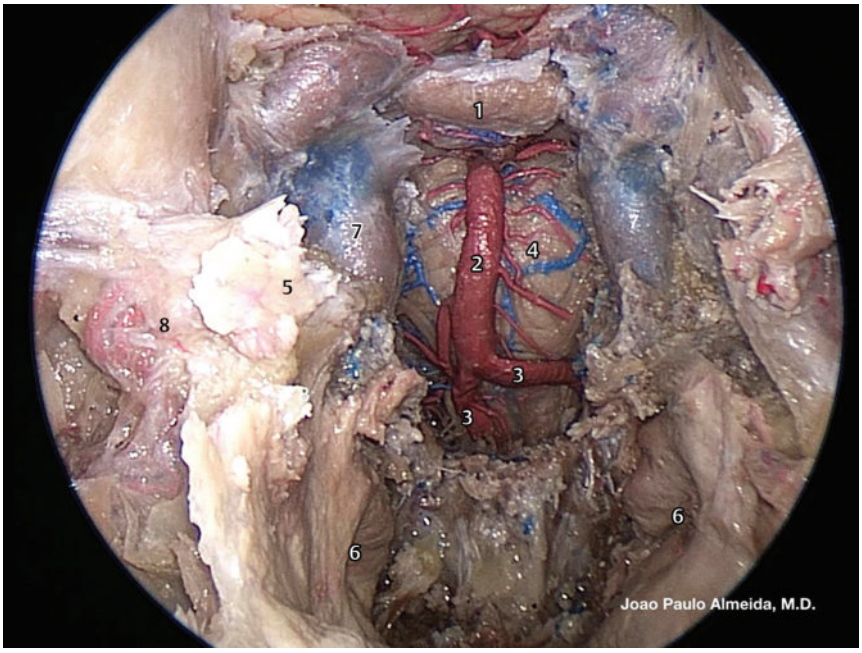


Fig. 17.8 Transclival approach and intradural exposure. 1, pituitary gland; 2, basilar artery; 3, vertebral artery; 4, pons; 5, vidian nerve; 6, eustachian tube; 7, paraclival carotids; 8, pterygopalatine fossa. (Copyright João Paulo Almeida, MD.)

chordomas with intradural and lateral extension, as the following case illustrates.

■ Case Presentation

A 49-year-old man presented with history of decreased libido, visual blurring, and decreased visual acuity. More recently, he also complained of headaches and nausea. On neurological examination, no cranial nerve or focal motor or sensory deficit was observed. As part of the investigation, the patient had an MRI scan which revealed a large clival lesion with intradural extension into the prepontine and interpeduncular cisterns with marked brainstem compression (**Fig. 17.9**).

■ Anatomical and Therapeutic Considerations

Based on the imaging findings, a clival chordoma was the most likely diagnosis. The size and location of the tumor, as well as its relation with the dura mater, cranial nerves, ICA, and vertebrobasilar complex should be assessed while planning the treatment. Because of the size and symptomatic nature, and in particular, the need for brainstem decompression, surgical intervention was necessary for this patient. The selection of the surgical approach must then take into consideration the goals of surgery (decompression, subtotal versus GTR), since some invasive chordomas are not amenable to radical resection.

Skull base chordomas are usually midline tumors that affect the clivus with varying patterns of extension in the skull base. Most are located medial to the petrous and paraclival segments of the ICA and cranial nerves V to XII that favor resection through a midline EEA. This approach also provides wide exposure for drilling of potentially affected portions of the clivus and sphenoid bones. While radical resection of midline chordomas may be achieved with this approach, those with lateral extension (such as those located lateral to the petroclival ICA junction and/or jugular foramen and those with invasion of the petrous bone) are more challenging and may not be amenable to radical resection via a pure endoscopic approach. Tumors with intradural invasion are also complex and have been classically associated with lower extent of resection and higher postoperative CSF leak rates, when compared to purely extradural chordomas.

Options for Approach

Anatomically, the clivus can be divided in three regions: upper, middle and lower clivus. Each portion has specific bone, cisternal, neural and vascular relationships. The upper clivus consists of the dorsum sellae and posterior clinoids and is located anterior to the interpeduncular cistern and upper prepontine cistern (**Fig. 17.10**). It is anatomically closely related to the oculomotor nerves, basilar apex, and cerebral peduncles of the midbrain. Additionally, the sella and pituitary gland are anterior to the upper clivus. If approached endoscopically, radical resection of chordomas in this location therefore requires the removal of the dorsum sellae, transposition of the pituitary gland (which can be performed via intradural, extradural, or interdural techniques) and control of venous bleeding originating from the cavernous or intercavernous sinuses.

The middle clivus is located medial to the petroclival junction of the ICA, at the level of the vidian canal and floor of the sphenoid sinus from an endonasal point of view (**Fig. 17.11**). The prepontine cistern, abducens nerve, pons, and basilar trunk are closely related to chordomas in this region. Endoscopic approaches to this region require extensive drilling of the sphenoid floor and control of bleeding from the basilar venous

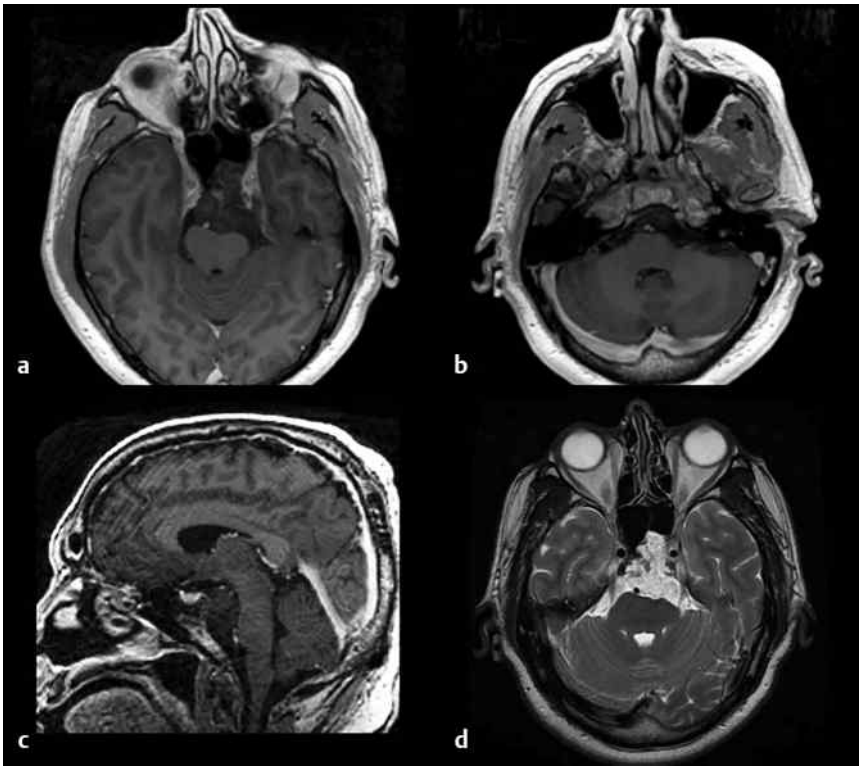


Fig. 17.9 Preoperative MRI. (a–c) A large clivus chordoma, mainly located at the middle clivus, but with extensions into the upper and lower clivus, was seen. **(d)** There were intra- and extradural components and a lateral extension around the left trigeminal nerve.

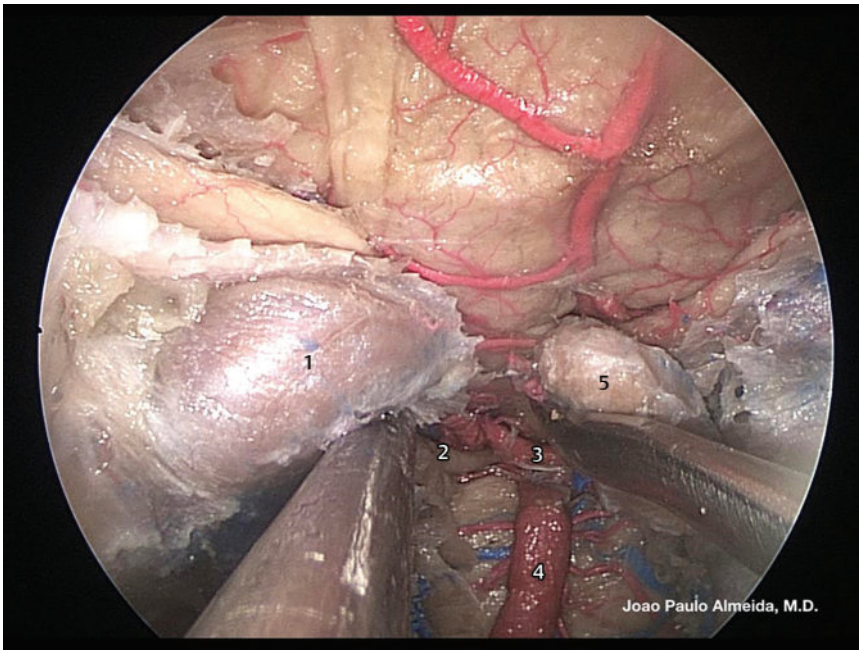


Fig. 17.10 Interpeduncular fossa exposure after pituitary transposition. 1, clinoid carotid; 2, right third nerve; 3, basilar tip; 4, basilar artery; 5, pituitary gland. (Copyright João Paulo Almeida, MD.)

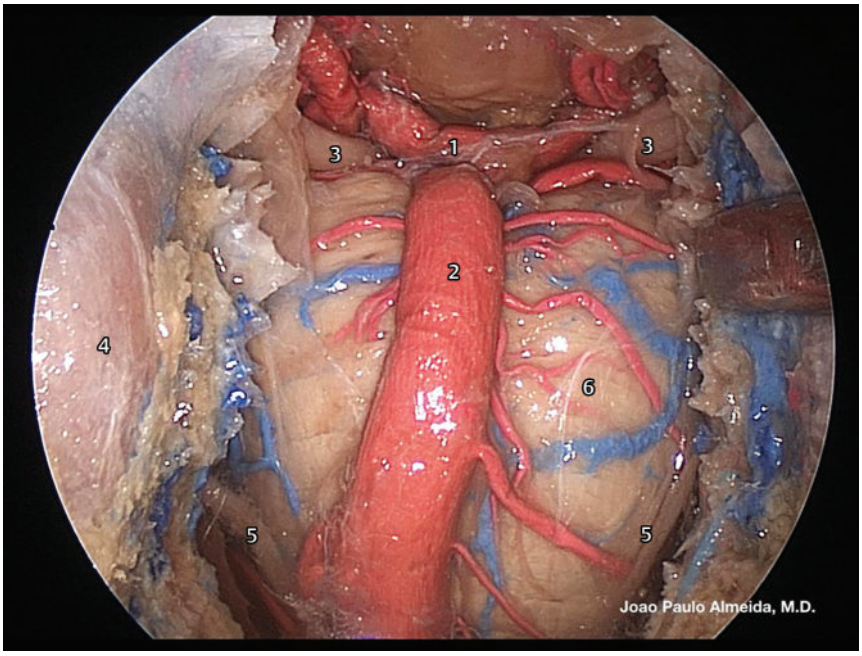


Fig. 17.11 Endoscopic view of the prepontine and interpeduncular cisterns. 1, basilar tip; 2, basilar artery; 3, third nerve; 4, paraclival carotid; 5, sixth nerve ; 6, brainstem. (Copyright João Paulo Almeida, MD.)

plexus. Tumors with lateral extensions toward the cavernous sinus, Meckel's cave, and foramen lacerum may be reachable with a transpterygoid extension of the transclival approach.

Again, from an endonasal point of view, the lower clivus is located posterior to the nasopharynx mucosa, nasopharyngeal fascia, longus capitis muscle, and rectus capitis muscle (**Fig. 17.12**). Therefore, exposure of this region requires mobilization of a large amount of soft tissue between the eustachian tubes. Midline drilling of the lower clivus will expose the premedullary cistern, the vertebrobasilar junction, the rootlets of the hypoglossal nerve, and the medulla. Tumor with lateral extension can be accessed by drilling the medial portion of the occipital condyle, which exposes the lateral part of the medulla and the region of the jugular foramen. Lesions with more lateral extensions, however, are less amenable to the endoscopic approach alone. Indeed, these lower clival chordomas with significant lateral extension have a less favorable prognosis.

Transcranial approaches are useful for resection of tumors with significant lateral extensions in the posterior fossa. Posterior petrosectomy may be suitable for resection of lesions located in the upper and middle clivus and extensions into the petrous bone and jugular foramen, while the far-lateral approach, or its variations, may be useful for resection of chordomas mainly located in the lower clivus with lateral extensions into the occipital condyles and foramen magnum.

A combination of endoscopic endonasal and transcranial approaches may also be useful for chordomas with these

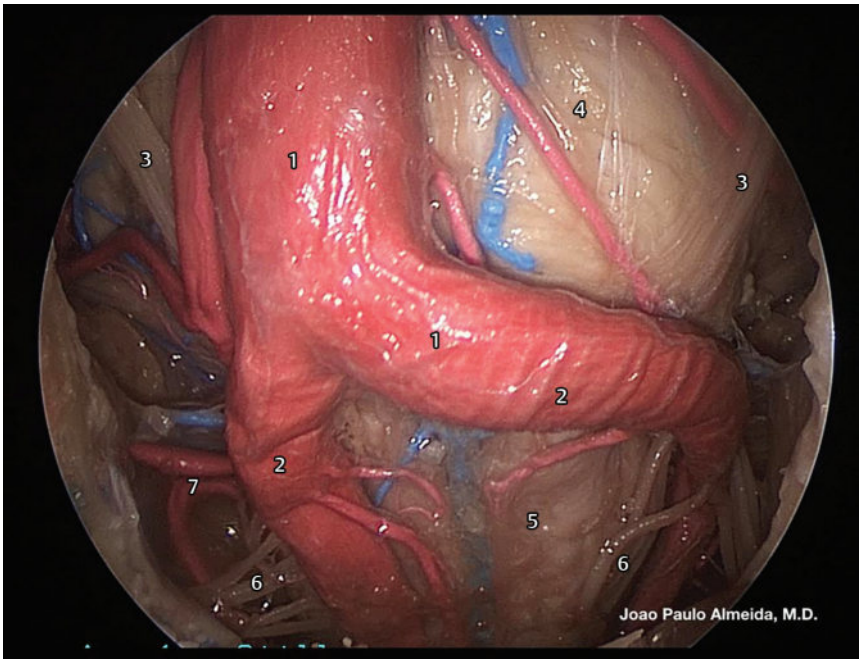


Fig. 17.12 Endoscopic intradural exposure—lower clivus. 1, basilar artery; 2, vertebral artery; 3, sixth nerve; 4, pons; 5, medulla; 6, twelfth nerve; 7, posterior inferior cerebellar artery. (Copyright João Paulo Almeida, MD.)

lateral extensions. That being said, we try to avoid combined approaches to minimize the morbidity associated with multiple approaches for management of a single tumor. Cases with bilateral extension pose a major challenge and may not be fully resectable even with a combination of different approaches. In such invasive cases, debulking of the tumor with decompression of the brainstem may be the most appropriate surgical goal. The ability to achieve a separation between chordoma and brainstem may allow delivery of the high dose of radiation while minimizing potential radiation side effects.

Approach of Choice

The chordoma in our case example was located primarily in the middle clivus with some extension to the upper and lower clivus. Although most of the tumor was in the midline, there was evidence of lateral extension with a component reaching the cerebellopontine angle and the left trigeminal nerve and VII–VIII nerve complex. Therefore, an endoscopic transclival approach with left transpterygoid extension was selected for resection of this tumor.

■ Description of the Technique

The patient was placed in supine position and intubated under general anesthesia. Neuronavigation was registered and electrophysiological monitoring was established for brainstem evoked potentials, somatosensory evoked potentials, as well as cranial nerves V to XII. The head of the patient was affixed to a Mayfield head holder, with the neck slightly flexed and rotated to the right. Prepping and draping were done in the usual fashion.

A 0-degree scope was used and the procedure was initiated with inspection of the right nasal cavity. In collaboration with our ear, nose, and throat (ENT) colleagues, a right middle turbinectomy and ethmoidectomy were then performed, followed by uncinectomy and a maxillary sinus antrostomy for subsequent flap storage. After identification of the sphenoidal ostium, a vascularized nasoseptal flap based on the right posterior septal artery was harvested and placed in the maxillary sinus for subsequent use at the end of the procedure. At this point, attention was directed to the left nasal cavity and a left middle turbinectomy, ethmoidectomy, and antrostomy were performed. Then, an additional nasoseptal flap was harvested from the left side and stored in the maxillary sinus (**Fig. 17.13a**).

Removal of the posterior septum and opening of the anterior part of sphenoid sinus were then performed. The floor of the sphenoid sinus was drilled down all the way back to the clival inlet. To achieve lateral exposure, a left transpterygoid approach was performed without ligation of the sphenopalatine artery, in order to preserve the ipsilateral flap (**Fig. 17.13b**). Drilling was done following the vidian nerve to the petroclival junction, maximizing the clival exposure. For exposure of the lower clivus, a needle-tipped monopolar was used to resect the nasopharyngeal mucosa in between the inferior turbinate and torus tubarius bilaterally with some resection of the longus capitis muscle (**Fig. 17.13c**). At this point, the clival tumor came into view, and we identified the carotid arteries bilaterally with Doppler ultrasound. The sphenoid floor was drilled flush with the clivus, further exposing the tumor (**Fig. 17.13d**).

The limits of drilling were the paraclival carotids laterally, the dura of the pituitary gland superiorly, and the foramen magnum inferiorly. The bone was removed over the pituitary fossa and the pituitary gland elevated superiorly to allow us to achieve a very high exposure including removal of the dorsum sellae. Ultimately, an excellent transclival exposure was achieved with a full view of the clivus from dorsum sellae to the anterior foramen magnum.

At this point, the dura was opened inferiorly and extended superiorly, first to the region of the tumor, and then further to reach the top of the clivus just below the sella (**Fig. 17.14a**). Radiating cuts were placed laterally. The tumor was then removed in a careful piecemeal fashion using microsurgical technique. The tumor was rather soft and responded readily to careful suction and curetting. Progressive tumor resection exposed the basilar artery, which had been shifted to the right side (**Fig. 17.14b**), and the tumor was removed from the central part of the brainstem. The resection of the superior component, as well as the

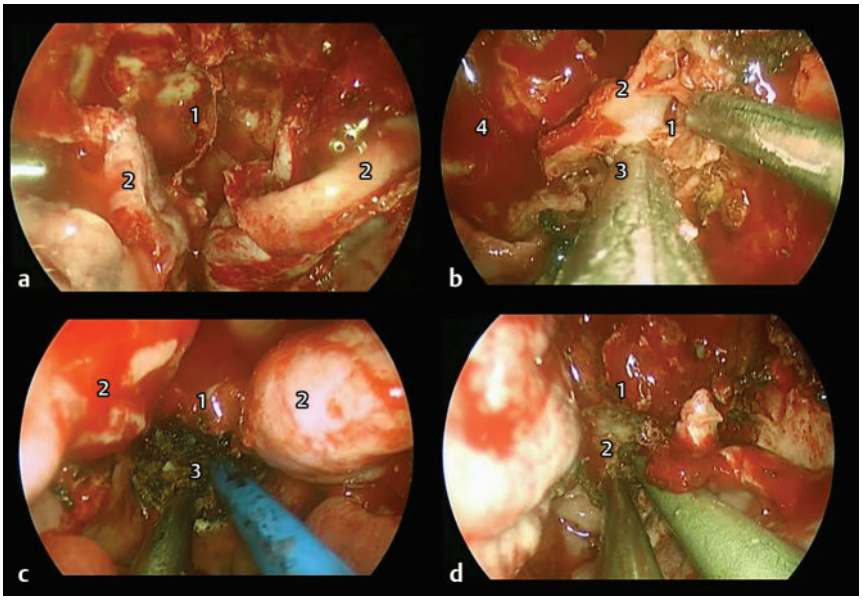


Fig. 17.13 Endoscopic endonasal approach with transpterygoid extension. **(a)** Sinonasal dissection. 1, sphenoid sinus; 2, vascularized flaps stored in the maxillary sinuses bilaterally. **(b)** Transpterygoid approach. 1, vidian canal; 2, pterygoid wedge; 3, medial pterygoid plate; 4, sphenoid sinus. **(c)** 1, floor of sphenoid sinus; 2, vascularized nasoseptal flaps; 3, dissection of nasopharynx mucosa and muscles. **(d)** 1, floor of sphenoid sinus; 2, lower clivus.

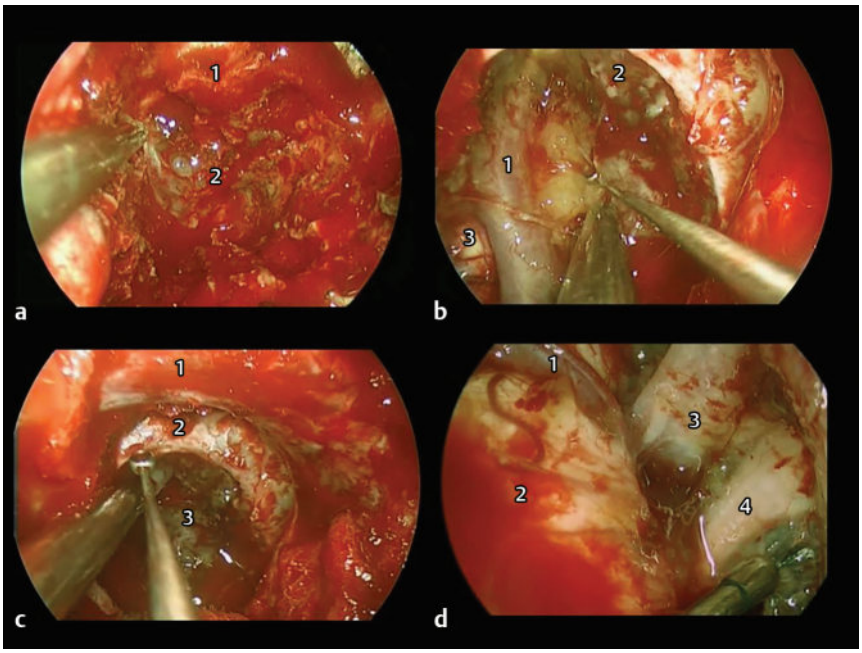


Fig. 17.14 Tumor resection. **(a)** Extradural component of the tumor. 1, sella; 2, tumor occupying the clivus. **(b–d)** Resection of intradural tumor. **(b)** 1, basilar artery; 2, tumor in the interpeduncular cistern; 3, pons. **(c)** 1, sella; 2, upper clivus dura; 3, tumor in the upper and middle clivus. **(d)** Exposure of the left cranial nerves V and VII–VIII and resection of the lateral tumor extension. 1, superior cerebellar artery; 2, pons; 3, nerve V; 4, VII–VIII complex.

part which extended toward the left, was facilitated by removal of the dorsum (**Fig. 17.14c**). The most lateral part of the tumor, close to the left trigeminal nerve, was carefully resected with the use of angled endoscope and angled suction (**Fig. 17.14d**). Ultimately, our intraoperative assessment was that a gross total removal had been achieved. At this point, the oculomotor nerve was seen superiorly in the interpeduncular cistern, and the trigeminal, abducens, facial, and vestibulocochlear nerves were all visualized (**Fig. 17.15**).

For skull base reconstruction, a multilayered repair was carried out. This involved initially harvesting of fascia lata and fat from the right thigh in the usual manner. The repair was carried out using Duragen inlay, followed by a fascia lata inlay, followed by a further fascia lata onlay. Then, the previously harvested nasoseptal flaps were placed over the bony defect, reinforced at the edges with Surgicel, followed by fat, and tissue adhesive. Finally, a balloon strut and Vaseline gauze packing were inserted to keep the reconstruction in place.

■ Aftercare

The postoperative course was uneventful, with no evidence of CSF leak or new neurological deficits. A CT scan was done 24 hours after surgery and demonstrated resection of the tumor with no complications. The Foley balloon and Vaseline gauzes were removed 4 days after surgery, and the patient was discharged on day 5 without complications.

An MRI was taken 1 month postoperatively and demonstrated a GTR of the tumor (**Fig. 17.16**). The final pathology report

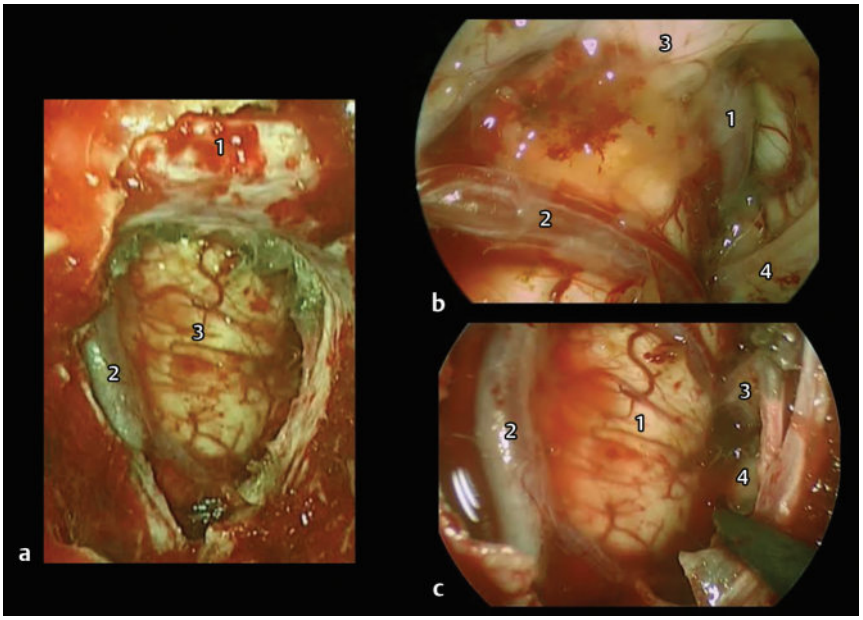


Fig. 17.15 Final intraoperative view after gross total resection. (a) 1, sella; 2, basilar artery; 3, pons. **(b, c)** 1, posterior cerebellar artery; 2, superior cerebellar artery; 3, oculomotor nerve; 4, trigeminal nerve.

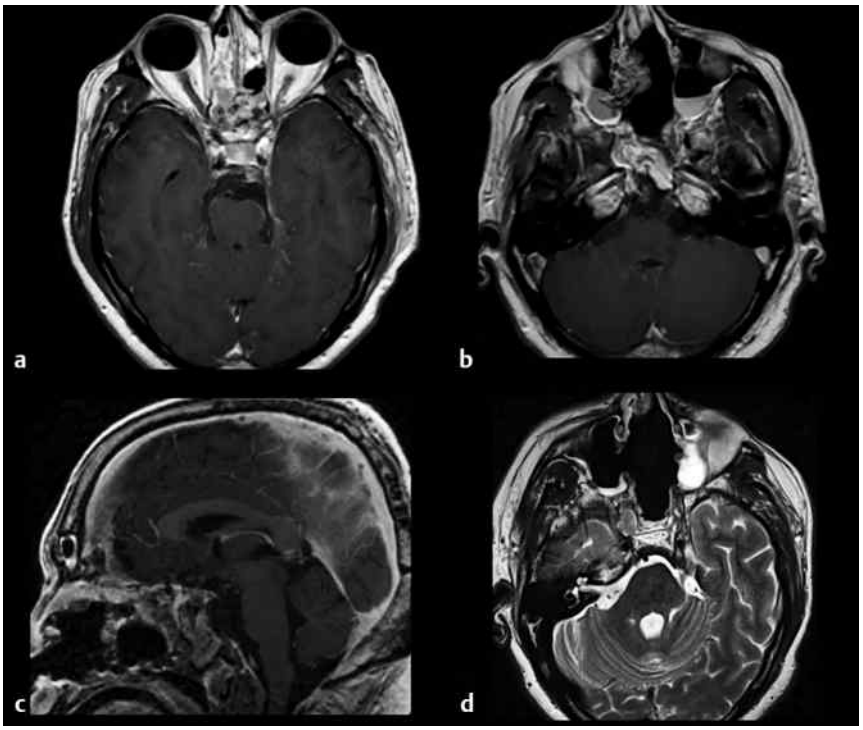


Fig. 17.16 Postoperative MRI. (a-d) These images demonstrated a gross total resection of the tumor and the multilayer reconstruction of the skull base with vascularized nasoseptal flaps and fascia lata.

confirmed the diagnosis of chordoma. Following the standard protocol at our Center for Chordomas, the patient was referred to radiation therapy, which was delivered by IMRT (78 Gy in 35 sessions).

■ Commentary

Endoscopic skull base surgery has had a significant positive impact, some would say revolutionized the treatment of chordomas, and is now a valuable tool in the armamentarium of skull base surgeons. It is a minimally invasive alternative to transcranial and transoral approaches to the clivus that has previously been considered as "gold standard" for the treatment of chordomas in this region. Successful treatment of skull base chordomas via endoscopic approaches require (1) deep knowledge of microsurgical and endoscopic anatomy of the skull base, (2) appropriate surgical tools and endoscopes, and (3) experienced ENT/head and neck and neurosurgery/skull base teams working in collaboration in a high-volume center. Midline extradural chordomas are excellent endoscopic targets, while tumors that present with significant lateral and intradural extensions are more challenging and may benefit from open or combined approaches.

Even in the hands of an experienced team of surgeons, the endoscopic techniques may not be universally applicable. It is

important to carefully review the anatomy of each case before selecting the surgical approach, as some cases are ill-suited for the endoscopic approach. Indeed, inappropriate case selection may lead to inadequate resection, intra- and postoperative complications. Although considered "minimally invasive," endoscopic techniques are associated with complications such as CSF leak, hypertensive pneumocephalus, meningitis, cranial nerve deficits, and stroke. Additionally, sinonasal complications in the endonasal corridor can significantly reduce the quality of life.

In the first section of this chapter, the authors presented a case example to show that open approaches remain important and necessary in the treatment of chordomas. In their patient, there was both significant intradural and deep lateral extension reaching the lateral aspect of the medulla, petrous bone/jugular foramen, and hypoglossal canal. These extensions would have significantly curtailed the resection via an EEA, and we would also have chosen a PLA. In contrast, in our case, the lateral extension was limited to the lateral margin of the trigeminal nerve, and this limitation made the endoscopic approach still applicable, albeit requiring angled endoscopes and instruments. Additionally, we encountered a soft tumor, the radical resection of which was feasible with the use of suction and microdissection. Had the tumor been more solid, we would likely not have had the same extent of resection.

In conclusion, we agree with the previous authors that both open and endoscopic techniques are required to treat chordomas effectively, and that skull base surgeons caring for these challenging patients should be prepared to offer the patient whichever approach has the best chance of a safe, maximal removal of the tumor. The choice should be based on the neuroanatomical features of the tumor, as well as objective analysis of the surgeons' experience, avoiding personal biases for or against any dogma.

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Part VI

Tumors Around the Petrous Bone

18 Petrotentorial Junction *231*

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18 Petrotentorial Junction

Shunchang Ma and Siviero Agazzi

Keywords: tentorium, brainstem compression, Jehovah's witness, suprameatal tubercle, meningioma

■ Case Presentation

A 36-year-old man presented to the emergency room with complaints of gait unsteadiness, poor coordination, headaches, transient left facial droop, tinnitus, and decreased hearing in the right ear. The physical examination was only remarkable for right lateral gaze nystagmus and decreased hearing on the right side. Facial movement and sensation were intact. His magnetic resonance imaging (MRI) revealed a 5.1 × 4.9 cm tumor in the posterior fossa, displacing the cerebellum and the brainstem. The patient was otherwise healthy, but of note, he was a Jehovah's witness. His hemoglobin was 14.7 g/dL.

Questions

1. What is the differential diagnosis of the MRI finding?
2. How does his religious belief impact on the surgical goals?
3. How does the tumor size influence the surgical approach?

■ Diagnosis and Assessment

The imaging data revealed an enhancing extra-axial tumor (**Fig. 18.1a**), with no erosion or enlargement of the internal auditory canal (IAC) (**Fig. 18.1b**). The tumor had a flat base against the petrous bone and was therefore mostly consistent with a meningioma. Other possible extra-axial lesions in this location include cranial nerve schwannoma, chondrosarcoma, and metastasis. All of these alternate diagnoses were unlikely based on radiographic appearance.

To properly set the surgical goal, these considerations were paramount: here was a young and healthy person, working full time until his admission to the hospital. Every consideration should be given to a complete resection of the tumor while minimizing neurological morbidity. Nevertheless, his religious convictions, precluding any kind of blood transfusion, significantly altered his risk profile and therefore our surgical strategy. Depending on the vascularity of the tumor, any operation on it might be aborted any time, and this compelled the surgeon to choose a strategy that places the most important part of the procedure near the beginning of the operation. In this case, decompression of the brainstem should be achieved as early as possible, accepting that the goal of complete resection might have to be abandoned or postponed.

■ Anatomical and Therapeutic Considerations

This tumor's base of insertion (and therefore its blood supply) appeared to be at the petrotentorial junction and superior petrosal sinus. Along the posterior petrous bone, it inserted at the level of the IAC, and extended anteriorly, stopping at the porus trigeminus, short of the clivus. There was no involvement of the Meckel cave or cavernous sinus (**Fig. 18.1a**). In the

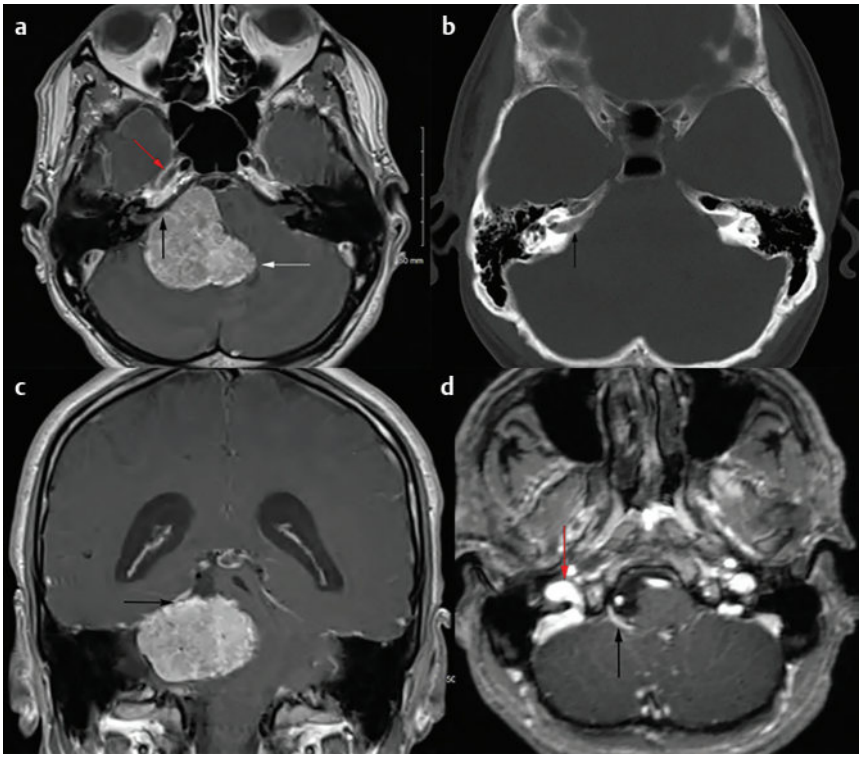


Fig. 18.1 Preoperative images. (a) Axial gadolinium-enhanced T1-weighted MR image. The superior *red arrow* shows the Meckel cave, the inferior *black arrow* pointing at the IAC, the medial *white arrow* shows the tumor compressing the brainstem. **(b)** Axial CT bone window showed no bony erosion and no enlargement of the IAC (*black arrow*). **(c)** Coronal CT showed the tumor location inferior to the tentorium; the *black arrow* shows that the tumor is not wrapping around the tentorial incisura. **(c)** Axial cut at the level of the jugular foramen showing no tumor. The superior *red arrow* pointing at the jugular foramen and the inferior *black arrow* pointing at the PICA which runs medially.

coronal plane (**Fig. 18.1c**), this tumor extended superior to the petrous bone by significantly displacing the tentorium upward, but there was no supratentorial extension. Caudally, the tumor extended to the level of the jugular foramen but did not involve the foramen itself (**Fig. 18.1d**).

The relationship of the tumor to the brainstem was very important for the proper selection of the approach. This tumor displaced the brainstem to the contralateral side and opened the cerebellopontine angle. It did not cover, or "cap," the brainstem posteriorly, whereas the right cerebellar hemisphere did "cap" the tumor and blocked the access to it from a posterior or posterolateral approach (**Fig. 18.2a**). Because of the patient's young age, the posterior fossa was "full," as all of the cisterns were compressed. This raised the concern that opening the cisterns may not achieve the usual cerebellar relaxation. On the T2 and FLAIR, the lack of signal change in the brain was a good sign that the separation between tumor and brain was preserved.

There was no tumor involvement of the basilar artery. The superior cerebellar artery coursed near the superior pole of the tumor (**Fig. 18.2b**) and the posterior inferior cerebellar artery near the lower pole of the tumor (**Fig. 18.2d**), but neither appeared engulfed. The anterior inferior cerebellar artery was much more difficult to clearly identify but was expected to run anteriorly near the equator of the tumor.

The oculomotor and trigeminal nerves ran superior to the tumor, with the latter displaced medially (**Fig. 18.1c,d**). The lower cranial nerves coursed along the inferior pole. The rest of the nerves were difficult to visualize. Until new technology makes this easier, one can make only an "educated guess" on the location of the seventh and eighth nerves based on tumor insertion. As this tumor inserted on the petrotentorial junction (i.e., superior to the IAC) and mostly anterior to the IAC, one can expect to find the facial nerve displaced inferiorly and posteriorly. This would create a wide working window between the nerves, but one must anticipate the less favorable situation where the seventh nerve would be located directly posterior to the tumor at its equator, putting it right in the middle of the working window.

Options for Approach

Two broad categories of approaches can be considered to access extra-axial tumors in this location: the retrosigmoid and transpetrosal approaches. Based on their relationship to the IAC, the transpetrosal approach can be further divided into posterior petrosal approaches (either retrolabyrinthine or translabyrinthine) and anterior petrosal approaches (sometimes referred to as Kawase's approach). Comparing the retrosigmoid and transpetrosal approaches, the latter are considered more of "skull base approaches," which means that they were designed to remove more bone, with the specific aim to decrease the need for retraction when accessing deep-seated tumors. The advantages of these approaches are undeniable, as a posterior transpetrosal approach would expose the posterior part of the tumor without requiring any retraction on the cerebellar hemisphere, and an anterior petrosal approach would expose the anterior part of the tumor without disturbing the brainstem until the end of the resection (**Fig. 18.3a**). These approaches offer a lateral trajectory toward the clivus, whereas the path of the retrosigmoid approach is posterolateral toward the center. This posterolateral path appeared to be blocked by the cerebellum in our patient (**Fig. 18.3b**).

Another advantage of the petrosal approaches is that, by operating lateral-to-medial, one would encounter the tumor's blood supply at its insertion (petrous bone and petrotentorial junction) before the tumor itself. This would theoretically decrease the tumor's vascularity at the outset and minimize blood loss during the operation. However, these "skull base"

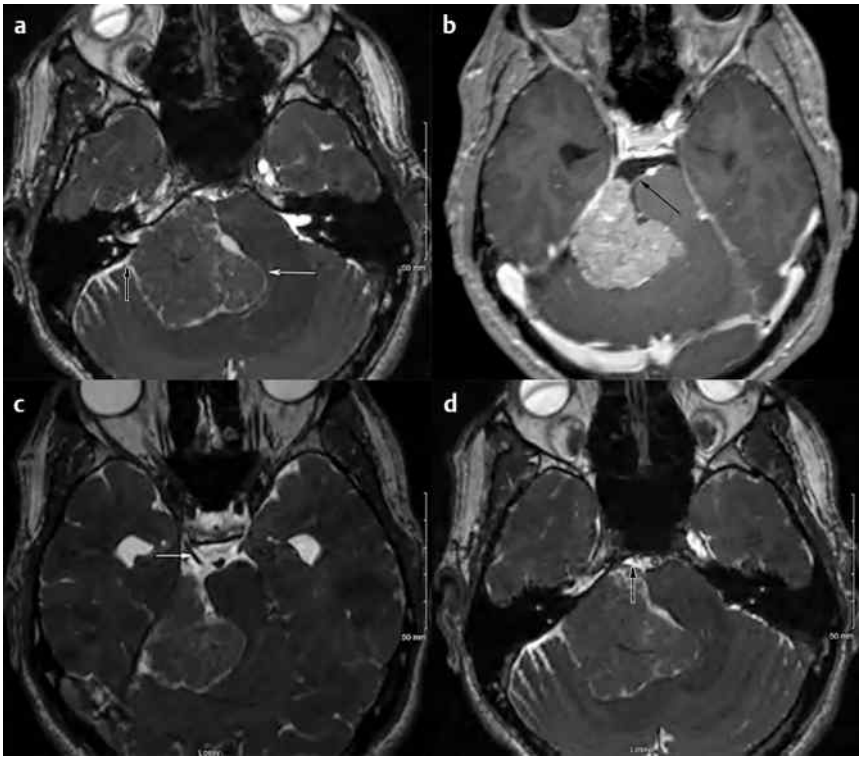


Fig. 18.2 Relationship with neighboring structures. (a) Axial T2 FIESTA MR image showed the tumor displaced the brainstem to the contralateral side. The *white arrow* demonstrates how the brainstem was open and was not capping the tumor posteriorly. The *black arrow* shows how the cerebellum wrapped around the tumor and capped it posteriorly, blocking access from a posterolateral approach. **(b)** The *black arrow* is pointing at the superior cerebellar artery which ran anterior and superior to the tumor. **(c)** FIESTA image showed the superior anterior part of the tumor. The *white arrow* shows the right third nerve which ran in the crural cistern on the way to the cavernous sinus. **(d)** FIESTA image showed the maximum cross-section of the tumor. The fifth nerve (*black arrow*) was anterior to the tumor and was displaced medially.

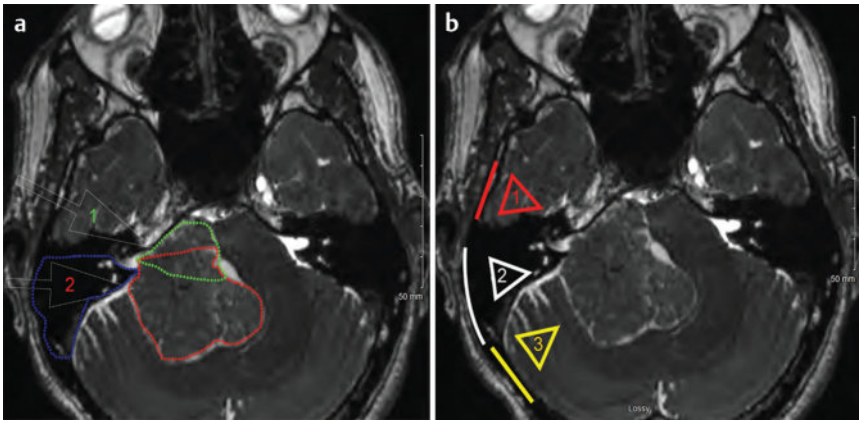


Fig. 18.3 Comparison of surgical exposures. (a) Axial T2 FIESTA MR image showed the lesion located in the posterior fossa. *Arrow 1* shows the exposure and trajectory of anterior petrosectomy approach. The *green dotted line* shows the expected tumor reachable with this approach. *Arrow 2* shows the exposure and trajectory of a posterior petrosectomy approach (retrolabyrinthine or translabyrinthine), and the *red dotted line* shows the expected tumor reachable with this approach. **(b)** *Triangle 1* in red shows, once again, the trajectory of anterior petrosectomy approach, the *red solid line* revealed the bone window. *Triangle 2* in white shows the trajectory of a posterior petrosectomy approach (retrolabyrinthine or translabyrinthine), and the *white solid line* shows the bone window. *Triangle 3* in yellow shows the trajectory of a retrosigmoid approach, the *yellow solid line* shows the bone window of the approach. Based solely on this figure, one might expect that significant retraction on the right cerebellum would be needed to expose the tumor. This was not the case during surgery.

approaches are not without downsides. One major disadvantage is the considerable time spent on the approach itself rather than on removing the tumor. Depending on the exact approach in question, the difference compared to a retrosigmoid craniotomy could range from a few to many hours. Furthermore, if the chosen transpetrosal approach involves decompressing a venous sinus, blood loss could be significant.

It is also worth noting that some limitations to the safe and complete removal of the tumor apply to both the retrosigmoid and the petrosal approaches: these are the relationship and degree of adherence between the tumor and the critical structures (brainstem, cranial nerves, and vessels). If the tumors were densely adherent to the brainstem, it would be just as difficult to resect via a petrosal as via a retrosigmoid approach.

Although far more "traditional" than the transpetrosal approaches, the retrosigmoid approach has undergone many refinements in recent years. This is in part due to the "trickle-down effect" of the skull base approaches, which has significantly improved our knowledge of anatomy at the cranial base. For example, craniotomies that used to just "open the foramen magnum" are now routinely expanded laterally to include the upper third of the occipital condyle and condylar fossa. As such, the "traditional" retrosigmoid approach has progressively become a more lateral approach that allows for an improved line-of-sight, parallel to the posterior petrous bone and flush with the posterior fossa skull base foramina. This, along with better anesthetic techniques for brain relaxation, good cerebrospinal fluid (CSF) drainage from the posterior fossa cisterns, and careful head positioning not to hinder venous outflow, has made it possible to remove most posterior fossa meningiomas without brain retraction. What appeared on the MRI, that the cerebellum completely blocked the posterolateral path to the tumor, is a static picture. The dynamic situation during surgery may be different. With good technique, the cerebellum can fall out of the way to safely expose the tumor without need for retractors.

Approach of Choice

Our final surgical strategy had to accommodate two competing factors: the patient's young age was the rationale for an attempt at total resection while the patient's categorical refusal of any blood transfusion required a strategy where the surgery could be interrupted if blood loss was too significant. The retrosigmoid approach was chosen mainly because it allowed us to start working on the tumor much earlier than with any transpetrosal approach. Expeditious tumor debulking and brainstem decompression were the primary goals, and must be achieved before hemodynamic issues can develop to truncate the procedure. We thought that the retrosigmoid approach gave us the best chance to achieve the primary goal, understanding that tumor devascularization would be the strongest determinant of the overall success. Applying the skull base *concepts* such as precise placement of the craniotomy to maximize exposure, early devascularization of the tumor at its insertion, and retractorless surgery, we were hopeful that the operation could go long enough for the achievement of the secondary goal, which was total resection. And by applying these concepts, we were in effect changing the retrosigmoid approach from a "traditional approach" to one which fits the criteria for being called a "skull base" approach.

Questions

1. How would you position the bone opening to maximize the exposure to the tumor?
2. Once the dura is opened, which cranial nerves would you expect to find? Which ones would you expect be hidden by the tumor?
3. How would you start the resection process once exposure is completed?

Description of the Technique

The patient was placed in the lateral position with the right side up. The head was kept relatively neutral and only slightly rotated toward the floor. The angle between the head and the shoulder was "opened" by flexing the head laterally toward the contralateral shoulder. All of these adjustments were very useful to improve the working trajectory, but the venous return from the contralateral sigmoid sinus and jugular vein should be preserved at all costs (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral

Craniotomy: retrosigmoid

Modifiers: none

A straight, retroauricular incision was made to expose the area of the asterion. For the best line-of-sight to this patient's meningioma, we planned to position the retrosigmoid craniotomy in a relatively lateral position, right up against the sigmoid sinus. In fact, the superior aspect of the craniotomy needs to expose the lower half of the transverse sinus and the lateral aspect of the craniotomy needs to expose the posterior half of the sigmoid sinus. We did not use burr holes. With a cutting burr, we first created a trough to form the superior and lateral edge of the craniotomy, and exposed the transverse and sigmoid sinuses. The craniotome was then used to complete the bone opening inferiorly and medially, always drilling *away* from the sinuses (see **Operative Setup**).

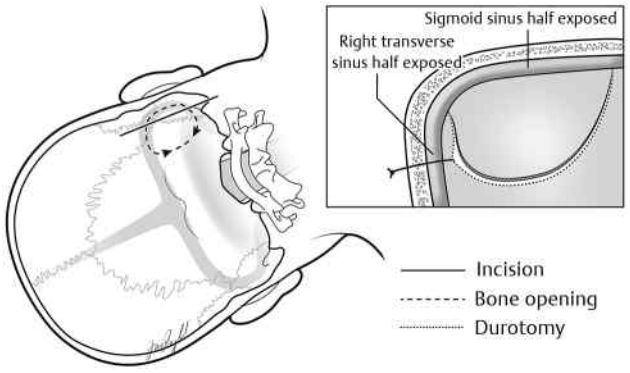
Operative Setup

Positioning: lateral, ipsilateral side up

Incision: linear, retroauricular

Bone Opening: retrosigmoid with partial exposure of transverse sinus/sigmoid sinus

Durotomy: C-shaped based on sigmoid sinus



A nearly semicircular dural incision was made, pedicled on the sigmoid sinus, with a few millimeters left below the transverse sinus to allow for an effective closure. That same dura along the transverse sinus would be retracted superiorly to increase exposure. The dura was opened inferiorly first. This allowed insertion of a cottonoid in the subdural space to reach the CSF cisterns very early on. A thin-tip bipolar was used to open the cistern magna to release CSF. Once the pressure was released, the rest of opening continued safely without cerebellar herniation. When the dura was fully opened, the cerebellar hemisphere spontaneously fell away from the petrous surface, exposing the posterior aspect of the tumor without any retraction.

To get oriented, we looked for the spinal accessory nerve, which, as always, was identified by its typical caudocranial trajectory right over the dura of the petrous bone. Following this to the jugular foramen led to discovery of the lower cranial nerves, and following these back to the brainstem, the choroid plexus of the foramen of Luschka was found. This exercise was important because, though hidden by the large tumor, the facial nerve is located about 5 mm superior to the choroid plexus. Also hidden at this time were the trigeminal and abducens nerve, which we expected to excavate much later in the operation near the petrotentorial junction.

Before any debulking has taken place, we were fortunately able to see the tumor insertion to the petrous bone and petrotentorial junction where the tumor's vascular supply originated. Given the importance of minimizing blood loss for this patient, we carefully proceeded with coagulating and severing this insertion. Disconnecting skull base meningiomas from their blood supply is best achieved by initially leaving a small amount of tumor on the insertion: some tissue that can be grabbed with a bipolar cautery to close off the feeding vessels. That thin carpet of tumor left on the tumor insertion would be addressed later. Failure to do so will make it impossible to have an effective and rapid hemostasis, as the feeding vessels will break and retract into the dural layers with no chance to quickly obliterate them. In this manner, while devascularizing the tumor, we left a cuff of tumor on the petrous bone dura and petrotentorial junction, with plans to return here later (**Fig. 18.4**).

Once the tumor had been fully devascularized, the ultrasonic aspirator was used to quickly debulk its core. It is critical to do this thoroughly, as debulking the core often allows the remainder of the tumor to progressively "deliver" itself into the operative field without the need to expand the operative window with retractors. Once sufficient work space was achieved after debulking, we mobilized the tumor away from the surrounding brain. Invariably, one can find some areas where the brain/tumor interface is preserved, and it is paramount to take advantage of these areas, and use them as starting points for the capsular dissection. As was often the case, we encountered some venous bleeding during the dissection, but it was important to use the bipolars only judiciously. Overzealous use of cautery can obscure the arachnoid layers between the tumor and brain and make capsular dissection much more challenging and dangerous. Once mobilized away from the brain, the tumor surface can then be cauterized (a minimum of 10 mm away from the brain).

With these maneuvers, we were able to remove the entire tumor and expose the trigeminal nerve from the brainstem to the trigeminal porus. As blood loss to this point was negligible,

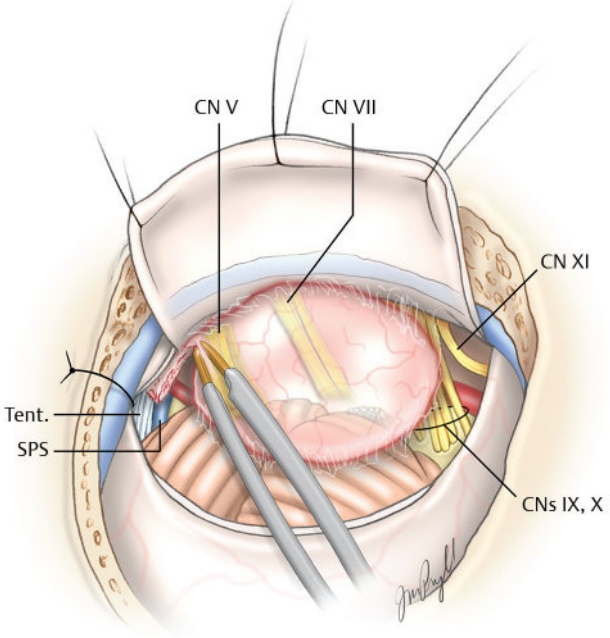


Fig. 18.4 Illustration of an intraoperative view. This surgical view illustrates the concept of devascularizing the meningioma from its attachment to the superior petrosal sinus (SPS) and petrotentorial junction at the very beginning of the surgery. A small cuff of tumor was purposely left on the insertion to allow a bloodless detachment of the meningioma from its insertion. This allowed the surgeon to have "something to coagulate" and avoid being left with holes in the sinus that are difficult to control when the bulk of the tumor still occupies the surgical space. Once the tumor was detached from its insertion and blood supply, the resection can continue with lesser bleeding. At the end of the procedure, when the tumor mass has been removed and there was ample space in the posterior fossa, the cuff of tumor left on the insertion can be fully removed from the sinus and the petrotentorial junction, as hemostasis would be easier to obtain with the wide working space available.

we made the decision to go after the tumor insertion, which had been previously left during the devascularization process. This included a cuff of tumor left on the superior petrosal sinus and the tentorium. As expected, removing those last pieces of tumor caused some brisk bleeding from the arterial feeders, the sinus and its draining veins, but those were easily controlled now that the space created by the tumor resection allowed us to maneuver instruments and hemostatic agents in the surgical field. The end result was a total resection with coagulation of the tentorium, petrosal dura, and superior petrosal sinus (**Fig. 18.5a, b**).

Having protected the dura from desiccation at the start of the operation, we were able to close the dura flap primarily using interrupted sutures. Onlay dural substitute and liquid sealant augmented the closure. After inspecting for any air cells which remained open, the bone flap was reattached, and the skin was closed in multiple layers. The blood loss was 250 cc.

Surgical Pearls

1. In removing a large meningioma, the first consideration should be addressing the blood supply. Preoperative embolization is always advocated but is rarely useful in skull base tumors. However, whether embolization is successful or not, the surgical strategy for removal of a skull base meningioma should involve attacking the base of insertion to devascularize the tumor early in the case. When doing so, it is important to realize that obtaining hemostasis from the tumor feeders right on the petrous bone surface, on any of the dural sinuses or up against the falx or the tentorium can be frustrating at best, and frequently ineffective and time consuming. Disconnecting skull base meningiomas from their blood supply is best achieved by initially leaving a small amount of tumor on the insertion: tissue that can be grabbed with a bipolar cautery to seal the feeding vessels. That thin carpet of tumor, left on the tumor insertion, is addressed later in the procedure, when the space left by the tumor resection allows for a better view and better reach of the feeding vessels without any brain retraction. The decision to remove the insertion or leave it in place depends on the overall strategy and goals of the surgery.
2. Once the tumor has been disconnected from its insertion, the resection must utilize the space provided by the tumor as a natural retraction system. The general concept is that the brain will re-expand as the tumor is debulked, and the access window to the petroclival region will slowly close. When compared to the retrosigmoid approach, the transpetrosal approach involves more bone removal and as such, there is less progressive closure of the surgical corridor as the tumor is removed. In the author's opinion, this concept alone makes the transpetrosal approaches the default approaches to these tumors, leaving the retrosigmoid approach as a shorter, less invasive but potentially more dangerous strategy. When using a retrosigmoid approach, tumor resection has to proceed in a very specific order in order to avoid a premature closure of the working window. The deepest part of the tumor (anteriorly and medially located) has to be debulked and removed first, leaving the more laterally and posteriorly located part to act as a natural retractor. The surgeon must resist the urge to place a retractor deep into the surgical field. That retractor will invariably end up pulling on the brainstem itself and cause major morbidity. History should be learned and not repeated; the unacceptable morbidity from brainstem retraction was one of the fundamental reasons for the development of the petrosal approaches.

■ Aftercare

The patient was extubated in the operating room and spent the first night in the intensive care unit. A few hours after the surgery, he was alert and oriented and his neurological examination was unchanged from preoperative stage; in particular,

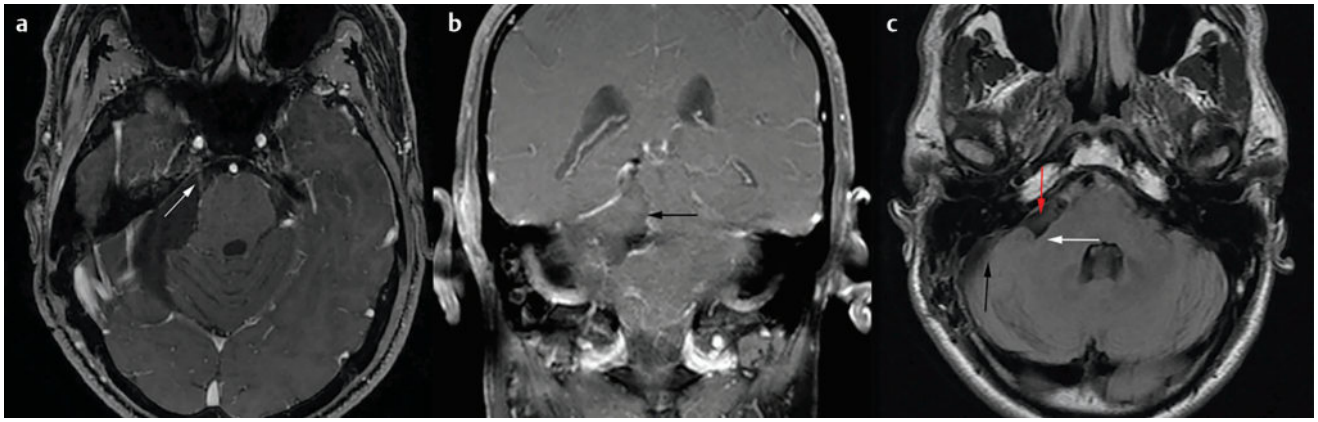


Fig. 18.5 Postoperative images. (a) Axial gadolinium-enhanced T1-weighted MR image at 3 months after the surgery. The cavity left after tumor resection could be observed. The fifth nerve (*white arrow*) was visible. No residual tumor was visualized. **(b)** Coronal gadolinium-enhanced T1-weighted MR image at 3 months after the surgery. The *black arrow* shows the resection cavity inferior to the tentorial incisura. No residual tumor was visualized. **(c)** The superior *red arrow* shows the cavity after tumor resection, the inferior *black arrow* points at the cerebellum which shows absence of atrophy, the medial *white arrow* shows the brainstem is in good condition with no encephalomalacia.

aside from decreased hearing on the right side, his cranial nerve function was intact, cerebellar testing was intact, and strength and sensation were also intact. He was mobilized on the first morning after the surgery and was able to ambulate independently. Venous thromboembolism prophylaxis was started 24 hours after the procedure.

A routine follow-up MRI was obtained 3 months after the procedure to establish a baseline and, because of the young age of the patient, long-term follow-up was planned. The MRI confirmed a gross total resection (**Fig. 18.5a, b**) with no signs of cerebellar atrophy or encephalomalacia (**Fig. 18.5c**).

■ Possible Complications and Associated Management

When a great resection is achieved and the patient comes out of surgery intact, one should not forget to thank the tumor. Even if we were not able to find any peer-reviewed study demonstrating this concept, every surgeon knows that tumor consistency and tumor adherence to critical structures remain major determinants of surgical outcome, irrespective of the surgical approach. When the tumor is not adherent to the brainstem or the cranial nerves, it can easily be “rolled into” the surgical field as the debulking progresses. Similarly, when basilar perforators or major arterial branches are not embedded in the tumor capsule, the tumor can be successfully mobilized and resected without causing either vessel injury or vasospasm.

Perspective

Anil Nanda and Devi Prasad Patra

When to take great risks; when to withdraw in the face of unexpected difficulties; whether to force an attempted enucleation of a pathologically favorable tumor to its completion with the prospect of an operative fatality, or to abandon the procedure short of completeness with the certainty that after months or years even greater risks may have to be faced at a subsequent session—all these take surgical judgement which is a matter of long experience.

—Harvey Cushing

■ Introduction

Tumors along the petrous apex are formidable lesions for at least two obvious reasons: first for its inherent tendency to involve multiple critical structures nearby, such as the brainstem and cranial nerves, and second for its deep-seated location, requiring complex surgical approaches for resection. Advancement of skull base techniques with better understanding of the anatomy has made the complete surgical excision of these extensive lesions possible, but surgical morbidity associated with these extensive skull base approaches is still a major concern. Additionally, the steep learning curve and long operative time have made these surgeries less appealing for neurosurgeons. With the introduction of radiosurgery, some of these lesions are now managed with subtotal safe resection and postoperative radiosurgery for residual tumors. The high degree of patient satisfaction and reduced morbidity associated with this strategy has significantly changed the treatment paradigm for these tumors.

As described by the authors in the previous section, a tumor in the petrotentorial region (with or without extension to the clivus) can be approached by a traditional retrosigmoid approach and a transpetrosal approach. The former involves a quicker craniotomy and early access to the tumor in the posterior cranial fossa. However, it comes with the disadvantages of limited access to the petroclival junction and increased working distance. Transpetrosal approaches provide direct and shorter routes of access, but require extensive bony work which delays access to the tumor during surgery. The most important factor that guides surgical approach is the tumor's relation to the tentorium and, if it escapes the confines of this dural barrier, where it extends beyond it. Other important factors include the degree of brainstem compression, presence or absence of hearing, and the functional status of the patient. An important consideration pertinent to the author's case is the potential need for blood transfusion in a patient who is a Jehovah's Witness. In cases like that, less extensive approaches like the retrosigmoid approach seem to be well-justified, allowing for early decompression of the brainstem with a shorter operative time. My personal opinion goes in line with the authors', in choosing the retrosigmoid approach for the case presented. Considering the location of the tumor, I would have chosen the same approach irrespective of the patient's characteristics. The judgment about choosing an approach becomes more complicated if the tumor penetrates the tentorium to involve the middle cranial fossa. We describe a similar patient with an extensive lesion along the petrous apex, which was managed with a multimodality approach.

■ Case Presentation

A 59-year-old woman presented with headache, difficulty walking, imbalance, and swallowing problems which were gradually progressing over a period of 9 months. Clinical examination showed impaired gag reflex, bilateral cerebellar signs, and spasticity in all the extremities. During objective testing, she failed her swallow test. She also had bilateral papilledema on funduscopic examination. An MRI of the brain revealed a large petroclival meningioma on the left side with supratentorial and middle fossa extensions. The patient had multiple comorbidities including congestive cardiac failure, hypothyroidism, anemia, diabetes, and hypertension.

■ Anatomical and Therapeutic Considerations

A detailed review of the MRI showed that the tumor had multicompartmental growth with the center around the left petrous apex (**Fig. 18.6**). The tumor extensions toward the posterior cranial fossa and the middle cranial fossa were nearly equal; however, the degree of brainstem compression was significant with displacement of the midbrain and pons toward the opposite side (**Fig. 18.6a–c**). It extended medially up to the midclivus and laterally up to the lateral third of the petrous bone. The tumor reached across the internal acoustic meatus, but did not extend into the meatus (**Fig. 18.6d**). Anteriorly it extended into the middle cranial fossa, displacing the inferomedial part of the temporal lobe laterally (**Fig. 18.6c**). The tumor also invaded into the cavernous sinus with expansion of its posterior part. Superiorly, the tumor extended through the tentorial hiatus between the free edge of tentorium and midbrain, displacing the medial temporal lobe structures superiorly (**Fig. 18.6b, c**). Inferiorly, the tumor reached the lower margin of pons and laterally up to the jugular foramen (**Fig. 18.6c**). The lower cranial nerves appeared free from the tumor without any gross displacement. Another important observation was the clear arachnoid plane between the tumor and brainstem and between the tumor and medial temporal lobe, seen as white arachnoid clefts in the T2-weighted images (**Fig. 18.6e**). There was associated ventricular enlargement limited to the lateral and third ventricles without any transependymal flow (**Fig. 18.6f**).

Although the tumor had significant multicompartmental growth, the symptoms of the patient best correlated with the posterior fossa component. The presence of the gait ataxia and bilateral cerebellar symptoms was possibly due to the compression of descending corticospinal tracts and crossing corticopontocerebellar tracts, respectively. The swallowing difficulty was related to a spastic pseudobulbar palsy of the pharyngeal muscle, which was best explained by the compression of descending corticobulbar fibers. The headache and papilledema were likely due to the hydrocephalus, as the triventriculomegaly pointed to tumor compression at the aqueduct. There was a relative lack of symptoms attributable to the middle cranial fossa extension (seizure or cognitive dysfunction),

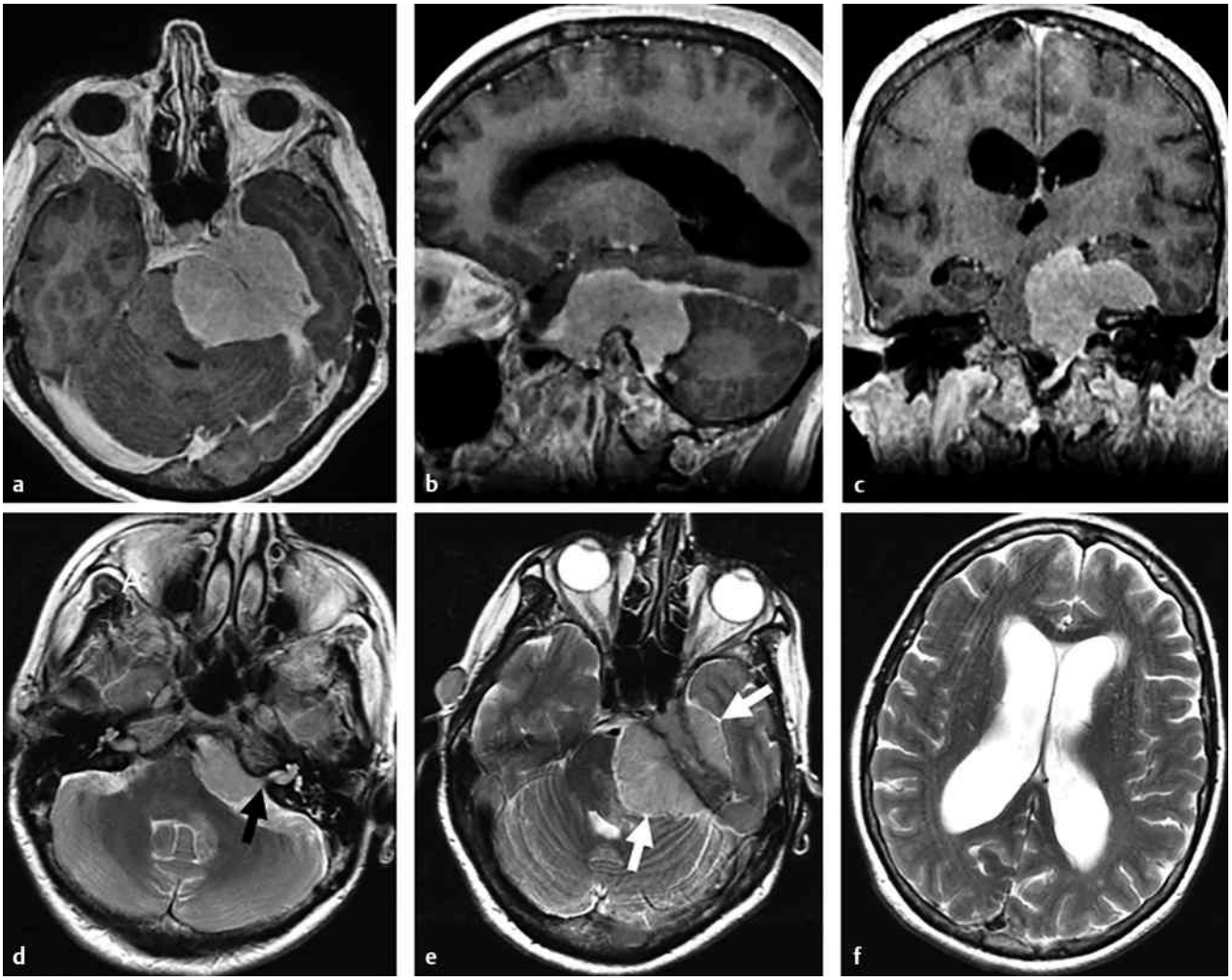


Fig. 18.6 Contrast-enhanced MRI showing the large multicompartmental tumor. **(a)** Axial, **(b)** sagittal, and **(c)** coronal sections showing extension of the tumor. **(d)** Note the tumor extending across the internal acoustic meatus (*black arrow*) but not extending into it. **(e)** Clear arachnoid plane between the tumor and the brainstem and the temporal lobe (*white arrows*). **(f)** Ventriculomegaly without evidence of transependymal flow.

cavernous sinus extension (ophthalmoplegia), supratentorial extension (fourth nerve paresis), or internal acoustic meatus encroachment (facial paresis or hearing problems). The overall clinical and radiological impression was a large petroclival meningioma with multicompartmental extensions, presenting with brainstem compression in the posterior fossa.

Options for Approach

Suboccipital Retrosigmoid Approach

This is a straightforward approach for this tumor as it is less time consuming and can tackle the posterior fossa component with relative ease. In the presence of a significant brainstem shift, this approach provides a great degree of freedom in the cerebellopontine angle. As mentioned earlier, this approach allows early brainstem decompression, but its major limitation is in accessing the midclivus region and the porus trigeminus.

With a modification called as "suprameatal approach," which involves drilling of the petrous apex medial and superior to the internal acoustic meatus, these tumor extensions, including the supratentorial components, can be reached (**Fig. 18.7**). Nevertheless, even a modified retrosigmoid approach does not provide adequate access to the middle cranial fossa, so tumors extending to the cavernous sinus and in front of the posterior clinoid are difficult to remove through this approach.

Posterior Transpetrosal Approach

Posterior transpetrosal approach would give a more direct and shorter route to the tumor and its attachments in the posterior cranial fossa. Many variations of the posterior transpetrosal approaches have been described including retrolabyrinthine, translabyrinthine, and transcochlear approaches which essentially involve increasing drilling of the mastoid and the middle ear. A retrolabyrinthine approach generates a presigmoid corridor with drilling of the mastoid apex and antrum. This gives early access to the tentorial and mastoid attachment of the tumor, and thereby allows early devascularization of the tumor. However, the medial exposure is limited and the compartments

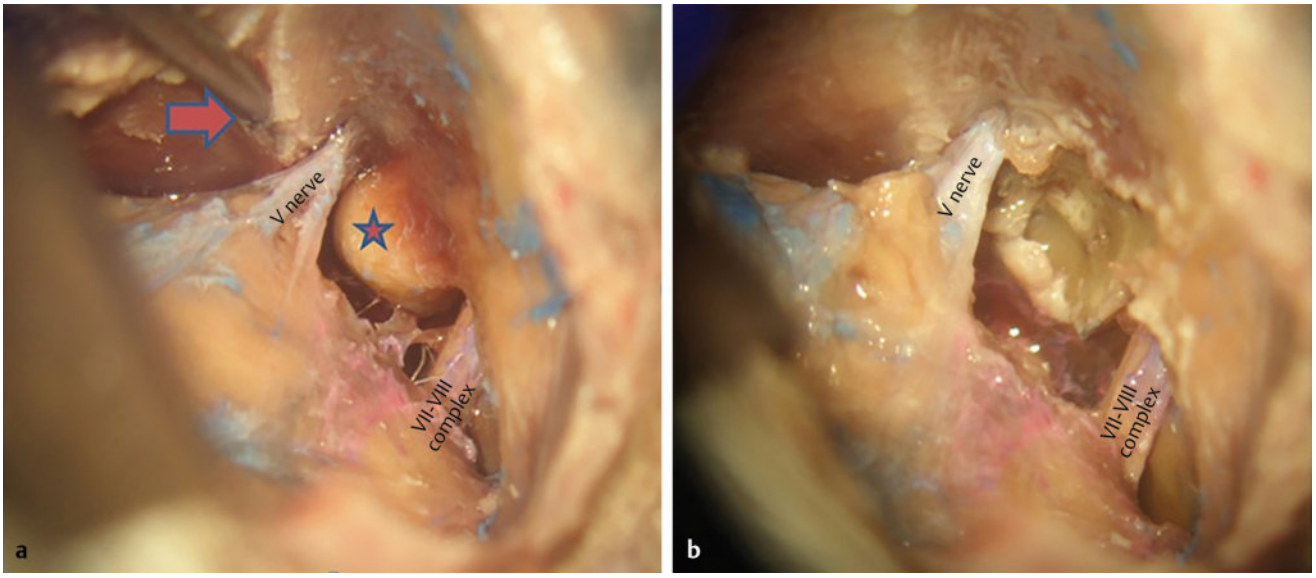


Fig. 18.7 Cadaveric dissection of the retrosigmoid approach with suprameatal drilling. (a) The retrosigmoid intradural suprameatal approach perspective, seen here with the identification of the suprameatal tubercle, which could be drilled to mobilize the trigeminal nerve and gain additional working space, to reach anterior and superior to the trigeminal nerve (crural and interpeduncular cisterns). Exposure of the cerebellopontine angle after retracting the cerebellum and incising the tentorium during the retrosigmoid transtentorial approach. The cranial nerve V can be seen in the upper part of the cerebellopontine angle entering into the Meckel cave. The *red arrow* points to a surgical probe in the area exposed after incising the tentorium. The *asterisk* indicates the suprameatal tubercle. **(b)** Area of exposure after drilling the suprameatal tubercle. Cranial nerve V can be seen traversing the superior portion of the cerebellopontine angle into the Meckel cave; nerve complex VII–VIII can be seen in the lower portion of the cerebellopontine angle. (Reproduced from Sharma M, Ambekar S, Guthikonda B, et al. A comparison between the Kawase and extended retrosigmoid approaches [retrosigmoid transtentorial and retrosigmoid intradural suprameatal approaches] for accessing the petroclival tumors. A cadaveric study. J Neurol Surg B Skull Base 2014; 75 (03):171–176.)

ventral to the brainstem are difficult to access. Translabyrinthine and transcochlear approaches involve progressive drilling of the bony labyrinth and the cochlea, respectively, offering more surgical freedom and medial exposure. However, these come with the price of losing vestibular and hearing function.

Anterior Transpetrosal Approach

This is also called an extended middle fossa approach, and it involves a middle fossa craniotomy and extradural drilling of the Kawase quadralateral, with a resulting access to the posterior cranial fossa from an anteromedial direction. This offers wide exposure of the petrous apex, middle fossa, posterior cavernous sinus, and upper clivus region, and is perhaps the best option for petrotentorial tumors with multicompartmental growth. A slight modification of this approach involves the extradural medial transposition of the gasserian ganglion that widens the bony drilling area and provides even wider access to the posterior cranial fossa inferiorly up to the internal acoustic meatus. Although quite appealing, this approach involves a moderate risk of nerve manipulation and temporal lobe retraction.

Approach of Choice

The important factors for our decision making were the patient symptoms, limited to the posterior fossa component, and her associated comorbidities. An anterior transpetrosal approach would have been our preferred approach, as it could give us the chance of complete excision with a single-stage operation. However, this approach has limited reach below the internal acoustic meatus and the possibility of a subtotal resection in the posterior fossa, where the brainstem compression was most symptomatic, made the option less attractive. Furthermore, the anterior transpetrosal approach requires a lot of time for drilling and in this patient with diabetes and cardiovascular problems, the risk of anesthesia complication was too high.

For obvious reasons, the posterior transpetrosal approaches were poor options in this patient with normal hearing and facial functions. Additionally, these approaches are equally as time consuming as the anterior transpetrosal approach with further risk of significant blood loss. Although the presigmoid retrolabyrinthine approach can handle the tumor vascularity in early phase, we would still have a blind spot at the dorsal midclivus, which needed definite decompression in this patient.

For these reasons, we considered the retrosigmoid approach as the primary approach for this tumor, since it gave us a good chance at adequate decompression of the posterior fossa component. The presence of the arachnoid plane between the tumor and brainstem, along with lack of involvement of the internal acoustic meatus, were favorable signs for an easy and safe resection. Similarly, the lack of T2 changes in the brainstem and absence of lower cranial nerve involvement were indicators for possible good symptomatic recovery from a simple brainstem decompression. However, given the patient's young age and natural life expectancy, the tumor extension in the middle fossa mandated treatment rather than simple observation. As such, we designed a multistage approach (three stages to be specific) for complete management of the tumor. In the first stage, we decompressed the posterior cranial fossa component using a retrosigmoid approach. In the second stage, we resected the middle cranial fossa component using the anterolateral

corridor (frontotemporal orbitozygomatic approach) leaving behind the cavernous sinus extension which was managed with radiosurgery in the third stage. The main philosophy behind our strategy was the maximal control of the tumor with the minimal possible morbidity.

Description of the Technique

1. First Stage (Suboccipital Retrosigmoid Approach)
   The steps involved in this approach were essentially the same as described in the first section of this chapter. The patient was placed in a lateral position in a Mayfield three-point fixation, with the head turned toward the opposite side. Frameless stereotactic system was used to map the lesion, sigmoid and transverse sinus. Neuromonitoring for the facial nerve was used. With a single S-shaped curvilinear incision just behind the mastoid bone, the skin flaps were retracted and the craniotomy was done to expose the transverse–sigmoid sinus junction. The dura was reflected laterally with tacking sutures. After initial CSF release from the cisterns, the tumor was evident as a large fleshy lesion (**Fig. 18.8a**). Initial superior dissection helped in decompression of a large part of the tumor with easy separation of the tumor from the trigeminal and cranial nerve VII–VIII complex (**Fig. 18.8b, c**). After decompression of the tumor from the brainstem, the posteroinferior part of the tumor was dissected from the lower cranial nerves and from the petrous attachment. A small medial part of the tumor was adherent to the brainstem and was intentionally left behind to avoid iatrogenic injury. After satisfactory resection of the posterior fossa component, hemostasis was achieved, dura was closed in watertight fashion, and the bone flap was replaced.
2. Second Surgery (Anterolateral Approach)
   Allowing sufficient time for the patient to fully rehabilitate from the first operations, this stage took place 6 months after the first. The patient was placed in a supine position with the head turned and tilted toward the opposite side with 30-degree neck extension. A curved incision from the midline down to the zygomatic arch in front of the tragus was made. An orbitozygomatic craniotomy was done using two burr holes, one placed near and above the sphenoid ridge and the second placed over the temporal bone. A one-piece bone flap was elevated and the bases of the sphenoid and temporal bone were drilled flat. The dura was opened in a C-shaped manner and was retracted anteriorly. The sylvian fissure was widely split and frontal and temporal lobes were separated. Transsylvian and subtemporal corridors provided a wide exposure of the tumor and its attachment to the base of temporal bone. After removal of the tumor from the middle cranial fossa base using ultrasonic aspirator, the tentorium and the tumor medial to it were exposed (**Fig. 18.8d**). The tentorium was divided, and the infratentorial part was also removed, taking care not to disturb the fourth cranial nerve. A small part of the tumor in the posteromedial edge of the tumor extending into the cavernous sinus was grossly adherent and was left behind. The tumor attachment to the base was coagulated. After confirming hemostasis, the dura was closed and bone flap was replaced.
3. Gamma Knife Radiosurgery
   Two months after the second surgery, the patient complained of left-sided facial pain and retro-orbital pain with slight numbness over the left side of forehead. The symptoms were attributable to the cavernous sinus component of the meningioma and therefore she was treated with radiosurgery. The prescription dose was 12 Gy at the 50% isodose line with a maximum dose of 24 Gy.

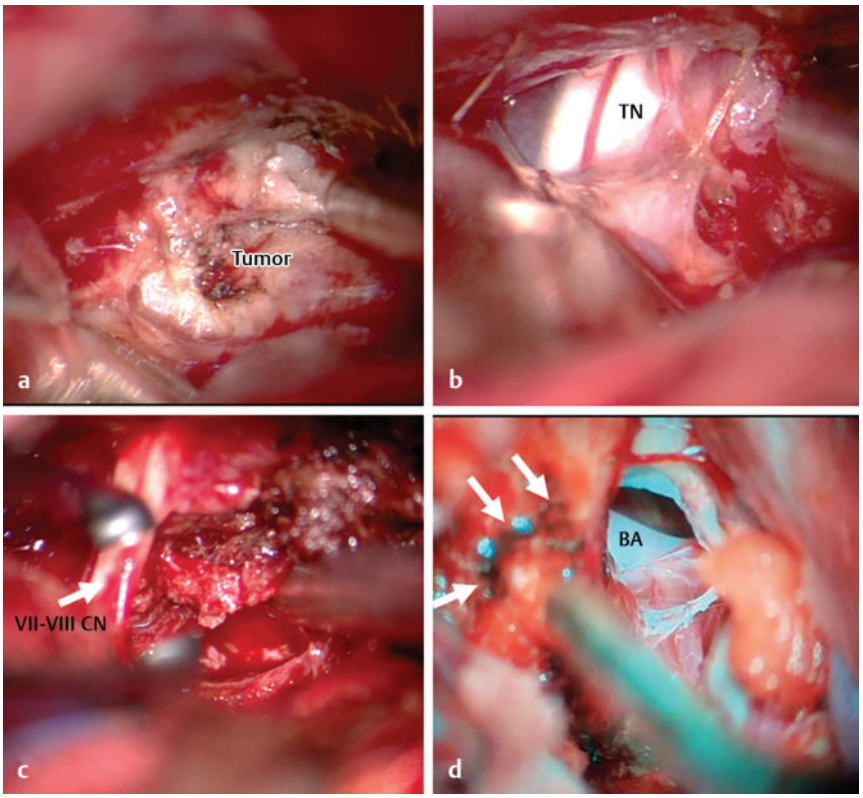


Fig. 18.8 Intraoperative pictures. (a) During the first operation via the retrosigmoid approach, a large fleshy tumor was seen. **(b)** The trigeminal nerve was visualized after resection of the superior pole of the tumor. **(c)** Dissection around the cranial nerve VII–VIII complex. **(d)** Intraoperative picture from second surgery showing the basilar trunk after complete resection of the tumor from middle fossa. Note the tumor attachment coagulated along the temporal base (*white arrows*). BA, basilar artery; TN, trigeminal nerve.

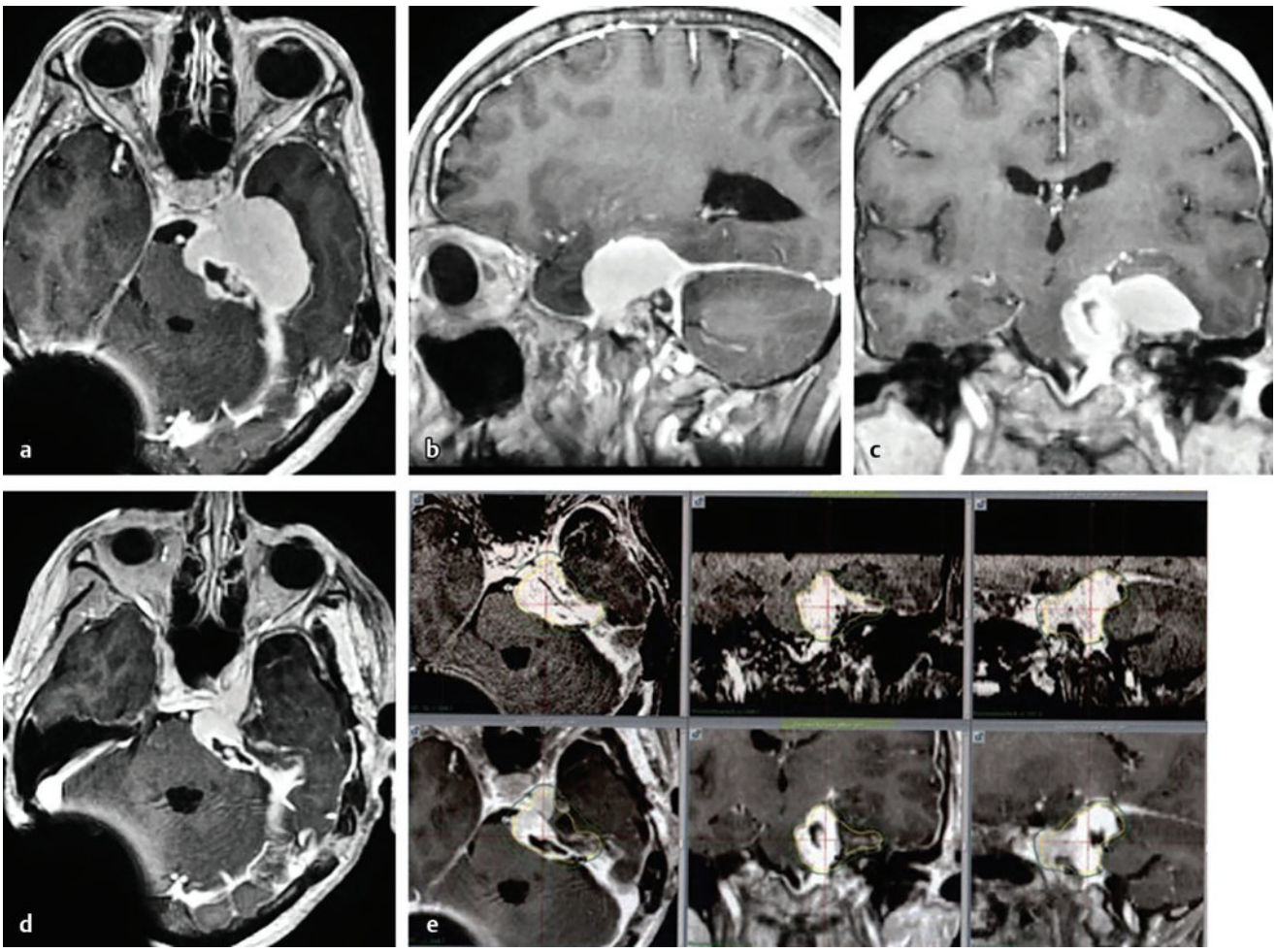


Fig. 18.9 Postoperative images. Upper panel shows the preoperative images from second surgery. **(a)** Axial, **(b)** sagittal, and **(c)** coronal contrast-enhanced MRI showing the residual tumor predominantly in the middle cranial base. Lower panel shows the postoperative images after the second surgery. **(d)** Postoperative axial contrast-enhanced MRI showing small residual tumor along the cavernous sinus and anterior brainstem. The residual lesion was subsequently treated with radiosurgery. **(e)** The radiosurgery plan.

■ Aftercare

After the first surgery, the patient continued to have headaches with nausea. She required ventriculoperitoneal shunt 3 days after the tumor resection, as her ventricular system failed to decompress despite removal of the posterior fossa tumor. After the shunt surgery, she was nearly asymptomatic. Her swallowing function improved completely as well as her balance. Her postoperative course after the second surgery was uneventful (**Fig. 18.9**). However, 2 months after the second surgery, she developed trigeminal symptoms from the cavernous sinus component of the tumor and received Gamma Knife radiosurgery. At 1-year follow-up after radiosurgery, she had mild facial pain which was controlled with medications.

■ Commentary

The petrotentorial tumor as described in the preceding section is a classic example of meningioma in the cerebellopontine angle. The situation is slightly complicated due to the patient's religious beliefs (patient being a Jehovah's Witness) restricting the option of blood transfusion if required during surgery. The retrosigmoid approach is the most viable option as appropriately described by the authors, since it provides quick and sufficient exposure to the tumor for brainstem decompression.

The major disadvantage of the retrosigmoid approach in our patient was the limited access to the medial retroclival and supratentorial components (along porus trigeminus). In the presence of significant brainstem shift, the retroclival area can be reached, but the operative corridor "naturally" created by the tumor must be maintained. The authors of the preceding section have described the preservation of the tumor capsule as a method to achieve this without retractors.

If our patient's baseline health status had allowed for longer surgery, a suprameatal drilling (above and medial to the internal acoustic meatus) would have created access to the porus trigeminus. But despite this modification, the middle fossa component of the tumor would have been difficult to reach through this corridor. Tumors with significant middle fossa component can be resected in a single-staged anterior petrosectomy approach if the posterior fossa component does not extend beyond the internal acoustic meatus inferiorly and laterally. However, this

approach extends the operative time even more, and patients with multiple comorbidities are probably better managed in a staged manner as described in this section.

The decision to "stage" an operation, or to use two separate approaches instead of one, must be tailored to the individual patient. A "second stage" not only increases the total operative time, but is also associated with increased risk from wound complications, scarring, and loss of arachnoid planes, as well as CSF leaks. As stated above, radiosurgery has dramatically altered the treatment paradigm for complex skull base tumors, changing the surgical goal from complete resection to safe, maximal resection in the majority of cases. It is therefore my general preference to perform single-stage operations in healthy patients to achieve this goal, and avoid the risks of a "second stage" aimed at completing the resection. It is particularly the combination of the patient's comorbidities and tumor anatomy, which led to the three-stage plan in this case example.

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19 Cerebellopontine Angle

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Keywords: vestibular schwannoma, hearing preservation, cochlea, facial nerve

■ Case Presentation

A 25-year-old man from an adjacent state came for another opinion regarding management options for a residual vestibular schwannoma and progressive symptoms. This extremely healthy young man presented elsewhere with, in retrospect, a 4-year history of mild left-sided hearing loss while serving in the Navy. This was not pursued and was not very bothersome. However, 6 months prior to the current presentation, he sought treatment because of sudden profound left-sided hearing loss, headaches, imbalance, and blurry vision. Evaluation at that time revealed papilledema and magnetic resonance imaging (MRI) showed a very large, left cerebellopontine angle (CPA) tumor consistent with a vestibular schwannoma measuring approximately 3.8 cm in posterior diameter with prominent hydrocephalus. He underwent a left retrosigmoid craniotomy and subtotal resection without complication. He awoke with no facial weakness or other new neurological deficits. A follow-up MRI scan the day after surgery revealed that the internal one-third of the tumor had been debulked and there was no evidence of hemorrhage or stroke (**Fig. 19.1a**). Ventriculomegaly persisted, but the patient reported improvement of his blurry vision (**Fig. 19.1b**). He was discharged 5 days after surgery with no new concerns. Further follow-up with his surgeon was not arranged.

Six months later, he presented to us with increasing visual complaints and persistent unsteadiness of gait. He noted when he would stand up from a seated position, his vision would "gray out" and he would be unable to see anything for between 5 and 7 seconds. He also noted persistent headache without nausea or vomiting, and difficulty walking long distances without holding onto something to steady himself. A follow-up MRI scan revealed the residual tumor now measured 4.3 cm, and there was edema in the adjacent brainstem and persistent hydrocephalus (**Fig. 19.2**).

Questions

1. How would you have approached the original tumor in this 25-year-old man with profound left-sided deafness, papilledema, and hydrocephalus?
2. What issues need to be addressed, and in what order, at this point when he presented with significant regrowth of tumor and persistent symptomatic hydrocephalus?
3. What surgical approach would you use to address the residual tumor?

■ Diagnosis and Assessment

Beyond the rapid regrowth of his tumor, we were very concerned about his visual symptoms. In particular, his untreated papilledema and hydrocephalus could lead to permanent visual loss. Therefore, the day after presentation, he was taken to the operating room for placement of a right parietal ventriculoperitoneal shunt with a Delta level 1 valve (Medtronic, Minneapolis, MN). His headaches resolved almost immediately and he reported improved balance and reduced blurry vision. A follow-up computed tomography (CT) scan 3 weeks later showed effective decompression of the ventricles.

■ Anatomical and Therapeutic Considerations

We had a detailed discussion regarding the best surgical approach to address the large residual tumor in this young patient with ipsilateral profound deafness and normal facial nerve function. Careful review of the MRI scan revealed there was a large component of tumor extending inferiorly almost all

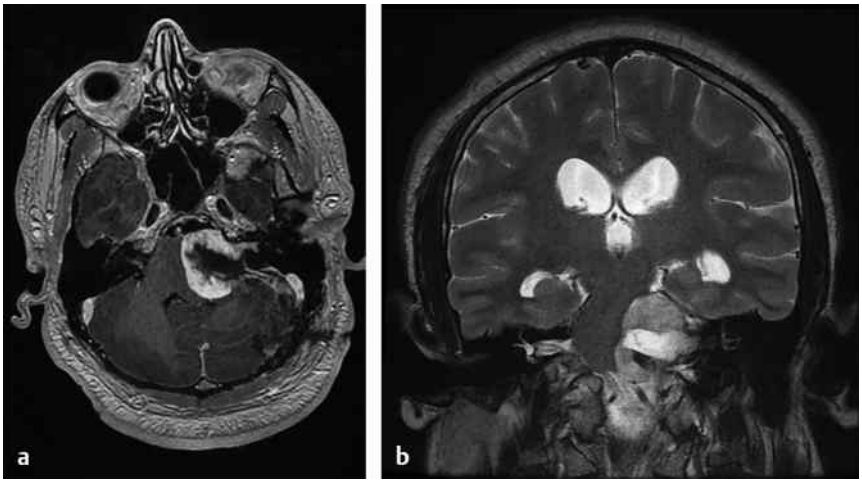


Fig. 19.1 Residual tumor. Immediate postoperative images after first resection. **(a)** Axial T1 MRI with gadolinium revealed the tumor had been internally debulked. **(b)** Coronal T2 imaging from the same study revealed persistent ventriculomegaly.

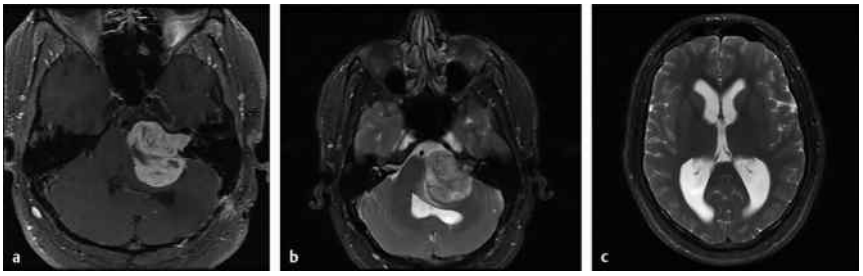


Fig. 19.2 Images 6 months after first resection. (a) Axial T1 with gadolinium 6 months following resection revealed a 4.3-cm residual tumor; **(b, c)** T2-weighted axial showed edema in the adjacent brainstem and persistent hydrocephalus.

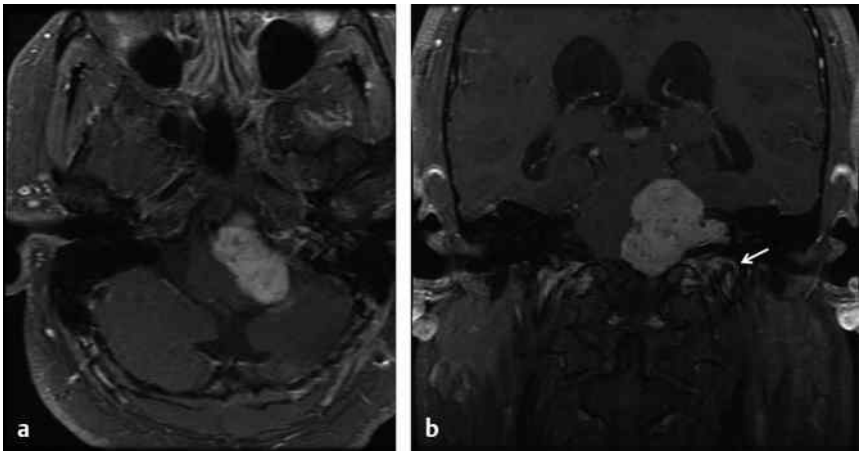


Fig. 19.3 (a) Axial T1 with gadolinium at the level of the medulla shows the tumor extended quite low and along the ventral medulla. **(b)** Coronal T1 with gadolinium revealed a prominent jugular bulb (*arrow*) and large residual tumor.

the way to the foramen magnum and extending ventral to the lower medulla (**Fig. 19.3a**).

Options for Approach

The three common surgical approaches for vestibular schwannoma are retrosigmoid, middle fossa, and translabyrinthine (**Fig. 19.4**). The retrosigmoid approaches provides a posterior-to-anterior trajectory to the tumor, and can be used for tumors both large and small. For the latter, there is a chance for hearing preservation with the retrosigmoid approach. The middle fossa approach offers the surgeon a superior-to-inferior view of the acoustic canal and is usually reserved for small or purely intracanalicular tumors in patients with intact hearing. The translabyrinthine approach reaches the tumor from a lateral direction and is most often used for patients with no salvageable hearing.

For this patient, one obvious option was to reopen the patient's prior retrosigmoid craniectomy and remove the tumor. Likely, the plane between the tumor and facial nerve was never dissected at the first operation. The retrosigmoid approach provides the largest view of the posterior fossa contents from tentorium to foramen magnum and is usually preferred for very large tumors at our center. The patient's prior retrosigmoid craniectomy had been closed with titanium mesh and reopening would mean operating through a scarred corridor.

In such a young patient with an aggressive-appearing tumor, we were equally concerned that any additional surgery should be curative with complete removal of the tumor. Such an operation, however, might lead to permanent facial weakness and require facial reanimation. With the fate of the facial nerve in mind, we considered a transpetrous approach, which provides access to the descending mastoid segment of the facial nerve that can be used for either a cable graft from the cisternal segment of VII or for a VII–XII anastomosis. Transpetrous approaches can provide limited or inadequate exposure if there is a large or high jugular bulb (**Fig. 19.3b**) or a contracted, small mastoid. In this patient, the jugular bulb was of average size and location, per our interpretation of the anatomical images. The most common transpetrous approach used for vestibular schwannoma is the translabyrinthine approach (**Fig. 19.4**). This necessarily sacrifices hearing since the inner ear is opened, but because this patient was already profoundly deaf, even before his initial retrosigmoid surgery, hearing was not a consideration. Because of the ventral extent of the tumor, we might need more exposure than the standard translabyrinthine approach provides and considered a transcochlear exposure. This involves closure of the external ear canal and either sacrifice or rerouting of the facial nerve which guarantees at least moderate, permanent facial nerve weakness. However, it also allows removal of the cochlea and the surrounding temporal bone, thus affording better exposure of the clival recess. The medial limit of the drilling is typically the inferior petrosal sinus and Dorello's canal containing the abducens nerve.

Approach of Choice

Ultimately, we decided on a translabyrinthine approach to avoid having to reopen and operate through the previously scarred retrosigmoid exposure. The translabyrinthine approach reduces the amount of retraction on his edematous cerebellum and brainstem and provides access for immediate facial reanimation if we were unable to preserve the facial nerve.

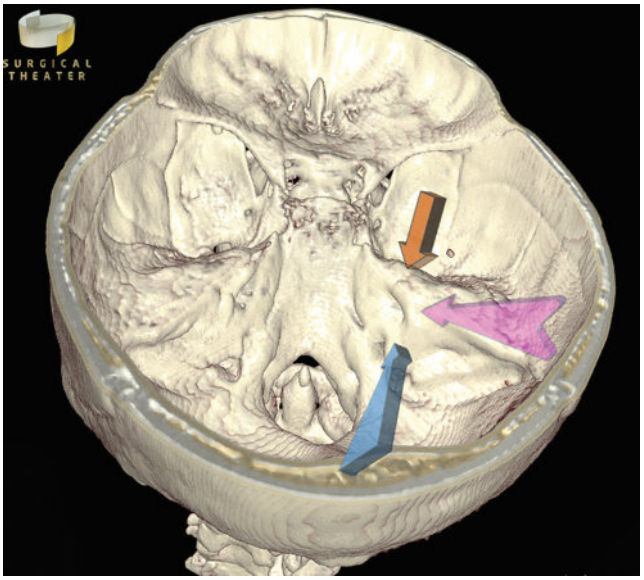


Fig. 19.4 Common surgical options for vestibular schwannoma. This view of the skull base shows the approach options to the right internal acoustic canal and cerebellopontine angle. The trajectory of the middle fossa approach is from top down (*brown arrow*), whereas that of the translabyrinthine approach is from lateral to medial (*pink arrow*). The retrosigmoid approach (*blue arrow*) reaches the internal acoustic canal from a posterior-to-anterior trajectory.

Description of the Technique

The patient was placed supine with the head rotated 45 degrees to the contralateral side. The head was placed in three-point pinion fixation. We prefer to use the Budde Halo retractor system (Integra Life Sciences, Plainsboro, New Jersey, NJ), which necessitates a rigid head holder. The previous incision was opened and extended anteriorly, and the soft tissue was mobilized anteriorly off the mastoid until the spine of Henle was reached, taking care not to breach the skin of the external ear canal (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral

Craniotomy: petrosectomy

Modifier: removal of bony labyrinthine

A standard wide mastoidectomy was performed (**Fig. 19.5a**). The tegmen was identified superiorly and the sigmoid sinus posteriorly, and both were completely decompressed of overlying bone. The mastoid tip air cells were removed. The Koerner septum was removed, and the antrum was defined. The horizontal semicircular canal (SCC) was identified and the short process of the incus was delineated (**Fig. 19.5b**). The vertical segment of the facial nerve was identified by visualization but was kept protected in a thin shell of bone. Then, the presigmoid dura was exposed from superior petrosal sinus superiorly, to the jugular bulb inferiorly (**Fig. 19.5c**). The endolymphatic duct and sac were identified and the cochlear aqueduct was opened. This maneuver often allows free egress of cerebrospinal fluid (CSF) with smaller tumors, which assists with relaxation of the intracranial contents. The bone posterior to the sigmoid sinus was also removed up to the patient's prior titanium mesh reconstruction (see **Operative Setup**).

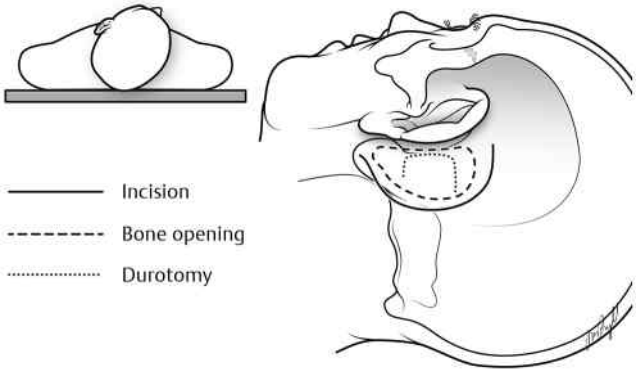
Operative Setup

Position: supine, head turned 45 degrees

Incision: retroauricular, curved

Bone Removal: translabyrinthine

Durotomy: "U-shaped" based on sigmoid sinus



A labyrinthectomy was then performed, removing the horizontal, posterior, then superior SCCs (**Fig. 19.5d**). The inferiormost aspect of the horizontal SCC was thinned but left intact, as the facial nerve runs along this and is most vulnerable during the drilling at this point. After the labyrinthectomy and the opening of the vestibule, the internal auditory canal (IAC) was identified and skeletonized about 270 degrees leaving a very thin rim of bone over the IAC dura (**Fig. 19.5e**). The drilling at the superior aspect of the IAC was done with copious irrigation and very carefully to safeguard the facial nerve. The thin bone fragments over the posterior fossa, middle fossa, and the IAC were carefully removed until the transverse crest separating the superior and inferior vestibular nerves was identified. After this, the dura was opened and the superior vestibular nerve was seen and the vertical crest (Bill's bar) underneath was palpated. After visualization and interrogation with the Prass probe, the superior vestibular nerve was deflected laterally and the facial nerve was identified.

The presigmoid dura was then opened just inferior along the course of the superior petrosal sinus with a parallel cut just superior to the jugular bulb. These incisions were connected by a vertical incision at the level of the porus of the IAC and the dura was reflected posteriorly over the sigmoid sinus. This nicely exposed the large tumor in the CPA (**Fig. 19.6**).

We found a very vascular and adherent vestibular schwannoma. After extensive internal debulking, we were able to establish a plane with the cerebellum and brainstem. Sacrificing the vestibulocochlear nerve allowed us to identify the facial nerve at the brainstem. We continued removing tumor in a piecemeal fashion and sharply dissect the tumor off the facial nerve. The plane was very unfavorable, and we noted decreased supramaximal electromyographic response as we worked. There remained tumor along the inferior ventral medulla which was hard to access and somewhat blocked by the course of the facial nerve.

After more than 9 hours of operative time we elected to stop the resection. By estimate, we had removed approximately 90%

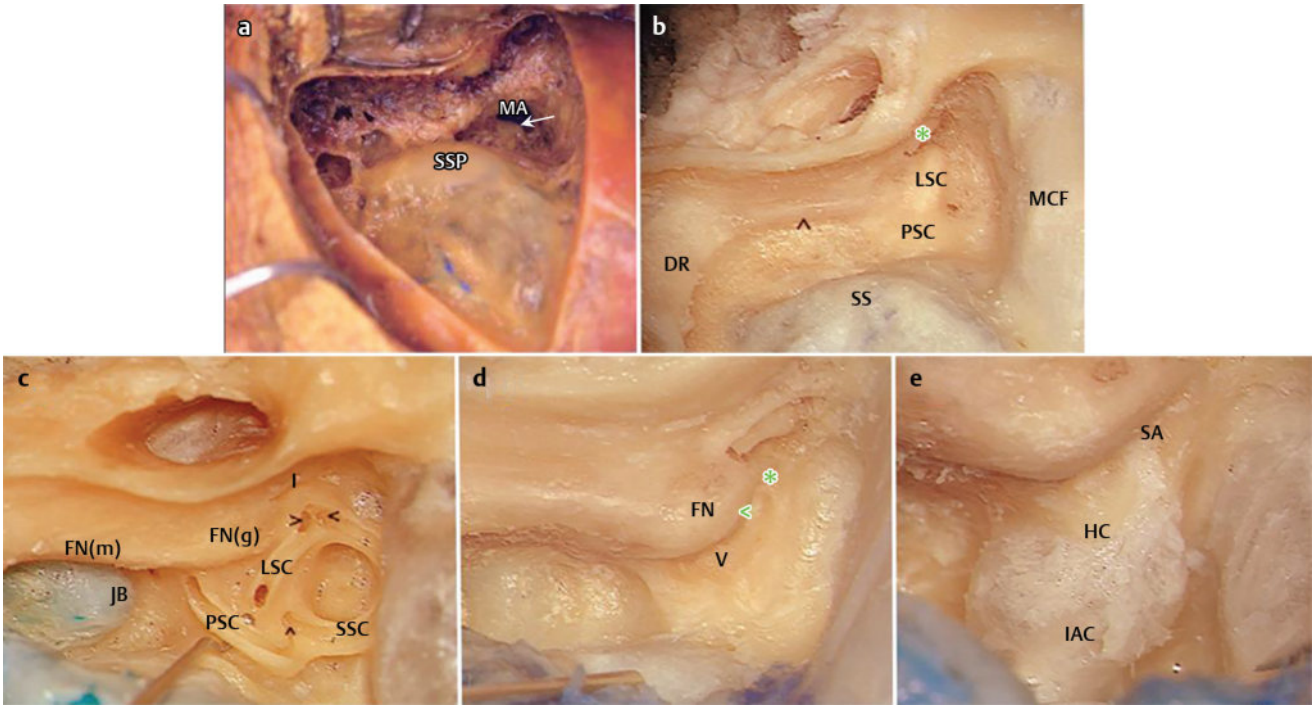


Fig. 19.5 Cadaveric dissection demonstrating the translabyrinthine approach on the left. (a) After a standard mastoidectomy, the first deep landmark encountered is the mastoid antrum (MA). The antrum is a large air cell. The hard bone of the lateral semicircular canal is seen protruding posteriorly into the mastoid antrum (*arrow*). **(b)** The digastric ridge points to the mastoid segment of the facial nerve (^). ∗, short process of the incus. **(c)** Relationship between the lateral (LSC) and posterior semicircular (PSC) canals with the facial nerve. Note that the sigmoid sinus runs medial to the facial nerve to become the jugular bulb (JB). **(d)** The canals have been drilled away and the vestibule (V) has been opened. Note the thin amount of bone (**<**) separating the genu of the facial nerve (FN) from the vestibule. ∗, the superior canal ampulla has been left as a landmark of the superior edge of the internal auditory canal. **(e)** The superior ampullary nerve (SA) is identified superiorly to the horizontal crest (HC) at the level of the fundus of the internal auditory canal (IAC). DR, digastric ridge; FN (g), second genu of the facial nerve; FN (m), mastoid segment of the facial nerve; I, short process of incus; LSC, lateral semicircular canal; MCF, middle cranial fossa; PSC, posterior semicircular canal; SS, sigmoid sinus; SSC, superior semicircular canal; SSP, sigmoid sinus plate.

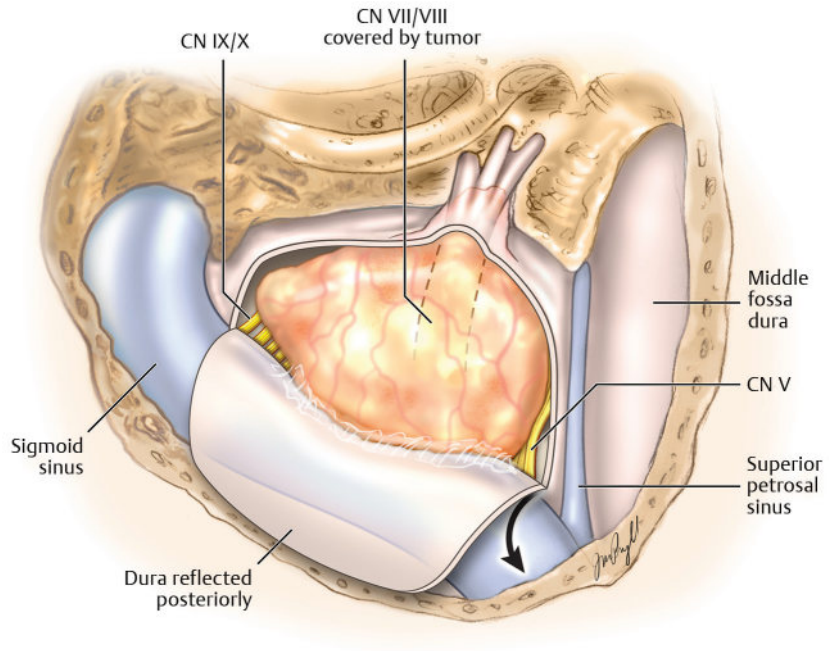


Fig. 19.6 Illustration of the intraoperative view. After the translabyrinthine approach, the dura was incised and reflected posteriorly. The large tumor is seen in the cerebellopontine angle, displacing the trigeminal nerve (cranial nerve V) superiorly, and the lower cranial nerves (cranial nerve IX/X) inferiorly. The brainstem and facial–vestibulocochlear nerve complex remained covered by the tumor at this stage of the operation.

of the tumor volume and the facial nerve was anatomically intact but with 86% drop off in proximal-to-distal supramaximal stimulation. We reasoned that while he would assuredly have facial weakness from the degree of facial nerve manipulation required to that point, there was a chance he could recover good facial nerve function and we had removed the great majority of the tumor. We decided it would likely be better to leave his facial nerve intact and treat the residual tumor with stereotactic radiosurgery (SRS). It was very difficult to manipulate the component of tumor along the ventral medulla and we were also concerned about putting his lower cranial nerves at risk.

Questions

1. Would you have pushed on with the resection to try and achieve a gross total resection (GTR), leaving the facial nerve anatomically intact but knowing further manipulation would almost certainly result in at least moderate permanent weakness?
2. Would you have sacrificed the facial nerve to achieve a GTR?
3. Would you have removed the titanium mesh placed at the first operation and reexplored the retrosigmoid approach to try and better access the ventral medulla region?

Surgical Pearls

1. The first operation provides the best chance for complete tumor resection and long-term tumor control. We almost always approach large tumors with the initial goal of GTR, and perform aggressive subtotal resection only when intraoperative factors point toward risking facial nerve function. In most first-time cases, the retrosigmoid craniotomy is our preferred approach for very large vestibular schwannomas, tumors with significant ventral extension anterior to the porus acusticus, and particularly tumors that extend inferior and anterior to the jugular foramen. In our hands, there is no difference in facial nerve outcome when comparing the translabyrinthine and retrosigmoid approaches in over 600 vestibular schwannoma surgeries. Furthermore, the retrosigmoid approach offers a better view of the lower cranial nerves and the trigeminal nerve as it approaches Meckel's cave. The exception to this may be the large tumor with disproportionately severe lateral-to-medial brainstem compression, for which the translabyrinthine approach provides a better angle for tumor resection. We recognize that this subject is heavily debated, and as with most things, the surgical team members should base their decision on what provides the patients with the best outcome in their personal experience.
2. For revision microsurgery after limited subtotal resection, we generally prefer to use an alternate surgical approach from the first operation. For example, in this case, we performed salvage surgery through the translabyrinthine corridor after the patient initially underwent a retrosigmoid approach at another hospital.
3. With a large tumor, it is often very difficult to accurately estimate true extent of resection without engaging the facial nerve plane both proximally at the brainstem and distally within the IAC. If the tumor was found to be adherent to the brainstem, then the hardest intraoperative decision is when to stop the resection to achieve the optimal balance between extent of resection and facial nerve outcome. This is mainly because there is no perfect intraoperative predictor of long-term facial nerve function. We use a combination of factors to optimally tailor extent of resection including patient age and expected longevity, patient wishes and willingness to accept any degree of permanent facial nerve weakness, intraoperative impression of facial nerve splay and tumor adherence, degree of neurotonic firing, increase in proximal minimum stimulation threshold, and proximal-to-distal supramaximal stimulation drop of greater than 70%.
4. In select cases where a patient presents with multiple recurrent aggressive disease, there may come a point where disease control trumps good facial nerve outcome. In the case example presented here, if the tumor continues to grow despite multiple rounds of surgeries and/or radiosurgery, we would likely recommend salvage surgery with a far-lateral approach to access the inferior/ventral disease. We would try to preserve the facial nerve but have a low threshold for facial nerve sacrifice and subsequent reinnervation, given his very young age and very aggressive disease.

■ Aftercare

The patient awoke from anesthesia with a House–Brackmann grade V facial function, with just a trace of movement in his upper face, but was otherwise well with no nystagmus, dysphagia, or dysphonia. A follow-up MRI scan on postoperative day 2 showed the expected residual tumor (**Fig. 19.7**). He was discharged home on postoperative day 5 with the plan for return 3 months following surgery with a new MRI scan and possible SRS.

■ Possible Complications and Associated Management

The patient contacted us 7 weeks following surgery complaining he was feeling more unsteady on his feet. He had no other focal neurological symptoms, no change in his House–Brackmann grade V facial function and specifically no new headache, fever, chills, weight loss, or drainage from the wound. His examination was stable with the exception of a more ataxic gait, and the wound was well-healed. It was therefore surprising that a follow-up MRI scan showed findings of a cerebellar abscess (**Fig. 19.8**). He returned to the operating room the following day when the prior translabyrinthine approach was reopened and the abdominal fat packing removed. The abscess cavity was debrided with the discovery of obvious purulent material. No new fat or other material was placed in the mastoid, and the wound was closed with monofilament suture. He tolerated this well with no change in

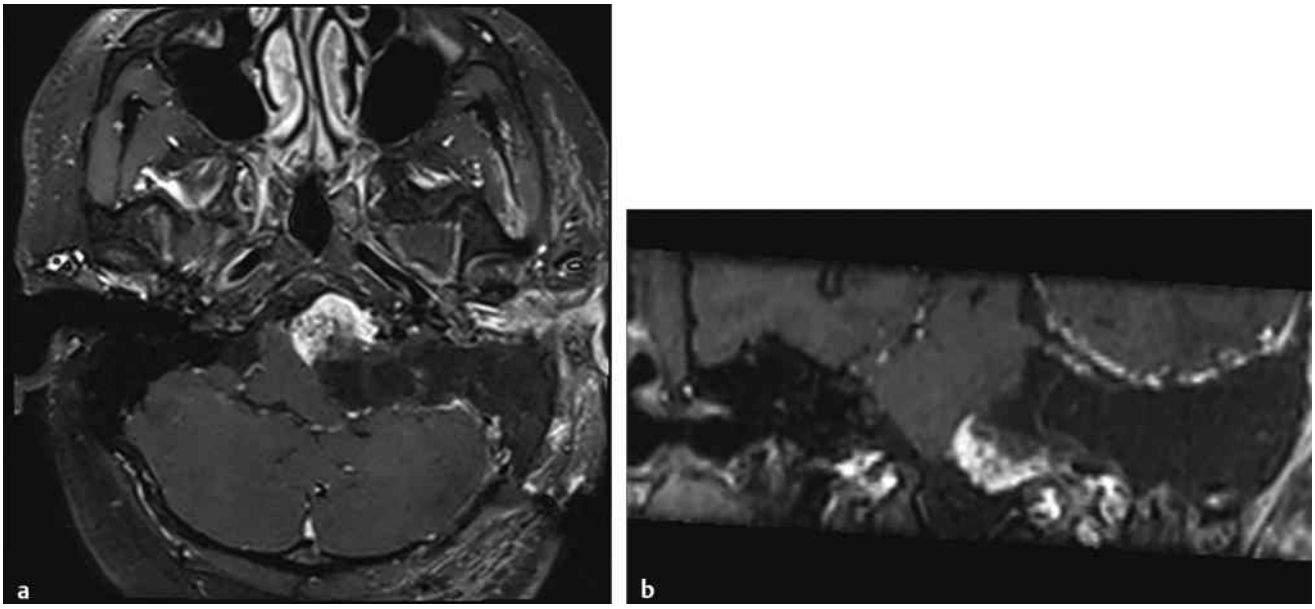


Fig. 19.7 Postoperative MRI after second resection. **(a)** Axial and **(b)** coronal T1-weighted images with gadolinium demonstrating residual tumor along the ventral medulla. These slices demonstrate the maximal extent of remaining tumor.

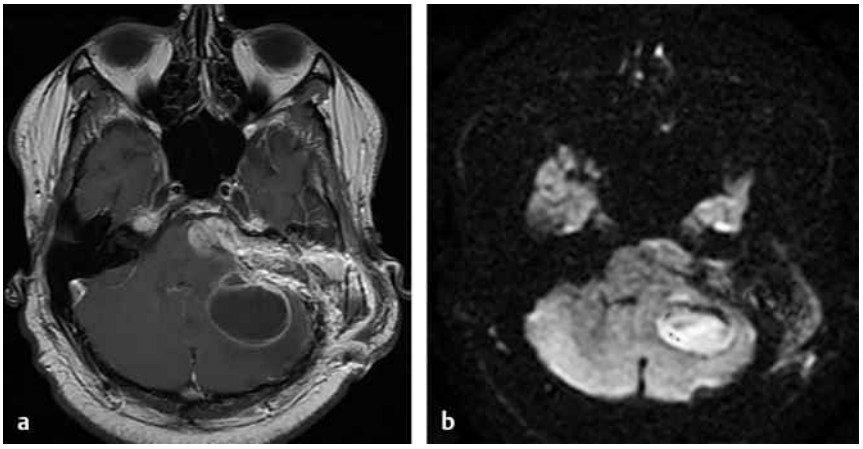


Fig. 19.8 Postoperative infection. **(a)** Axial T1-weighted MRI scan 7 weeks after subtotal resection of large residual vestibular schwannoma shows a new peripherally enhancing cystic structure in the cerebellum. **(b)** Diffusion-weighted imaging showed bright signal consistent with cerebellar abscess.

his examination. Cultures from this operation grew methicillin-sensitive *Staphylococcus aureus*, and the patient was treated with a 6-week course of intravenous Nafcillin, with marked improvement of his gait ataxia.

He was seen in planned follow-up 3 months following the repeat resection of his vestibular schwannoma and at the conclusion of his antibiotic course. Another follow-up MRI showed resolution of his abscess but concern that the residual tumor may be larger compared to the immediate postoperative MRI scan (**Fig. 19.9**). His facial nerve function had improved to House–Brackmann grade II and he felt well.

As such, 4 months after his repeat subtotal resection, his residual tumor was treated with radiosurgery (SRS) using the Gamma Knife (Elekta AB, Stockholm, Sweden). A volume of 9.17 cc was treated with a marginal dose of 13 Gy and a maximum dose of 26 Gy. The patient has done well with stable House–Brackmann grade II facial function and no new complaints. Most recent follow-up shows some additional residual enlargement. We continue to follow him with biannual MRI scans.

Obviously, if his tumor is not controlled with SRS, we are going to face very difficult decision making. Almost certainly, any additional surgical resection must lead to a complete tumor removal (GTR) with the necessary sacrifice of facial nerve and then reanimation. In our experience, this radical approach can be quite difficult without significant risk to the other regional cranial nerves, and vasculature to the cerebellum and brainstem. Some centers advocate repeat SRS if it is not effective initially, but given our liberal dose plan and large treatment volume (nearly 10 cc), we would be reluctant to radiate this tumor a second time in this young man.

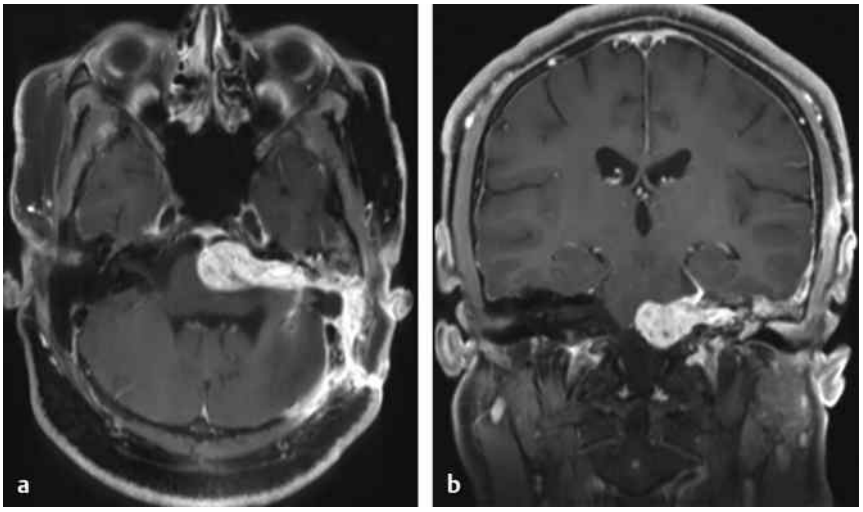


Fig. 19.9 Resolution of infection. **(a)** Axial and **(b)** coronal T1-weighted post-gadolinium MRI scan obtained 3.5 months postoperatively showed the residual tumor looked slightly larger, accounting for expected progression of postoperative changes.

Perspective

Gillian L. Harrison, J. Thomas Roland Jr., and John G. Golfinos

■ Introduction

When choosing an approach to large vestibular schwannomas, often defined as greater than 3 cm in the extrameatal space, a multitude of factors must be considered. In addition to those which have been repeatedly discussed in this book, preoperative hearing status and facial nerve function must be specifically taken into account for vestibular schwannomas. Tumor recurrence or progression after prior treatment, such as described in the previous section, adds an additional level of difficulty and increases the risk of postoperative complications. Surgeon experience and comfort additionally play a significant, though difficult to quantify, role in surgical management.

In general, larger tumors are associated with higher rates of residual or recurrence and higher rates of postoperative hearing loss, facial nerve dysfunction, and complication. Many groups have reported their surgical outcomes for large vestibular schwannoma. Most studies, however, are single-center retrospective reviews spanning long time periods, thus limiting generalization. A clear advantage of the retrosigmoid approach in comparison to the translabyrinthine approach is the possibility of hearing preservation, although achieving this in patients with large vestibular schwannoma is an uncommon feat. Complicating the discussion of surgical approach selection for large vestibular schwannomas is the neurosurgical "elephant in the room": Most neurosurgeons are more comfortable with the retrosigmoid approach, as it is used not only for vestibular schwannomas, but also for a variety of other posterior fossa pathologies from meningiomas to epidermoid tumors.

The retrosigmoid approach is often hailed for its "panoramic" view of the CPA, even though the visualization of tumor extending to the lateral end of the IAC is extremely limited, possibly resulting in higher rates of residual tumor. Furthermore, many neurosurgeons were exposed in their training to cerebellar retractors created specifically for the retrosigmoid approach, such as the Janetta retractor. Cerebellar retraction may result in permanent ataxia and ipsilateral dysmetria, and has been associated with higher rates of postoperative CSF leak and headache. For this reason, we do not use retractors, even when performing a retrosigmoid approach, allowing cisternal CSF drainage and gravity to do the job of displacing the cerebellar hemisphere.

While the translabyrinthine approach eliminates the possibility of hearing preservation, it is a true "keyhole" skull base approach. As such, it too provides a wide view of the CPA through a smaller cranial opening than the retrosigmoid approach. More importantly, the translabyrinthine approach provides direct and full access to the lateral aspects of the tumor and the fundus of the IAC, including the vertical and horizontal crests. One of the gifts of the translabyrinthine approach is early identification of the facial nerve at the fundus and a greater ability to identify the correct plane between the facial nerve more anteriorly and the superior vestibular nerve just posterior to it. Just as important as the access to the lateral end of the facial nerve is the more anterior angle of approach provided to the medial end of the facial nerve at the brainstem. Because this is a presigmoid approach, the surgical approach angle to the facial nerve root entry zone, the vestibulocochlear nerve, and the flocculus is more anterior and therefore more direct (**Fig. 19.10**). For large tumors, this more-direct trajectory is often critical in accessing the medial tumor that burrows into the foramen of Luschka or knuckles into the middle cerebellar peduncle itself.

Because of the many advantages elucidated above, the translabyrinthine approach is the preferred approach at our institution for vestibular schwannoma greater than 3 cm.

■ Case Presentation 1

A 53-year-old man presented to another hospital with a 2-year history of right-sided hearing loss followed by progressively worsening balance, difficulty with handwriting, and intermittent slurred speech. The preoperative audiogram revealed profound asymmetric sensorineural hearing loss on the right side. MRI revealed a 4-cm, partially cystic right vestibular schwannoma with early hydrocephalus (**Fig. 19.11**). He underwent a right retrosigmoid craniectomy for planned subtotal resection at the outside institution, with an intention to treat the residual with radiosurgery.

His postoperative course was uncomplicated, although he lost hearing on the right. The residual tumor was larger than expected, and it progressed after just 6 months (**Fig. 19.12**). On presentation to our institution, the patient reported intermittent headaches, persistent imbalance, and right facial numbness and tingling. On neurological examination, he had absent hearing, diminished facial sensation in V1–V3 on the right, and

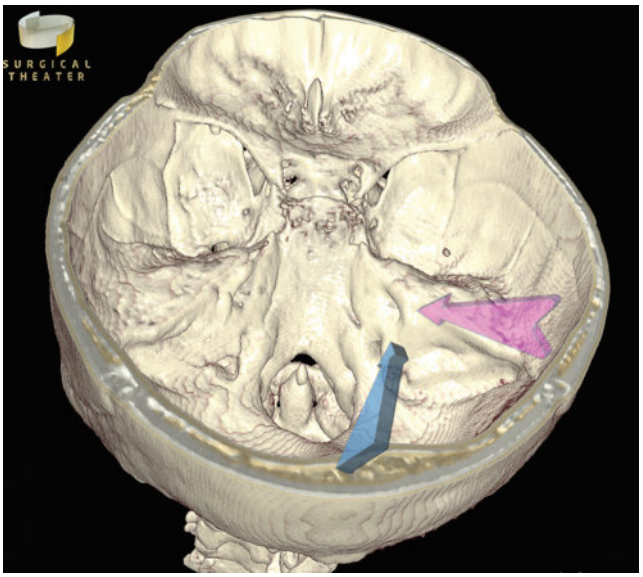


Fig. 19.10 Comparing the surgical trajectory of the retrosigmoid and translabyrinthine approaches. Because the translabyrinthine (*pink arrow*) is a "presigmoid" approach, the surgeon is placed more anteriorly, and has a more lateral-to-medial line-of-sight. This trajectory is more direct to medial structures such as the takeoff of the facial nerve at the brainstem. The trajectory of the retrosigmoid approach (*blue arrow*) is posterior-to-anterior.

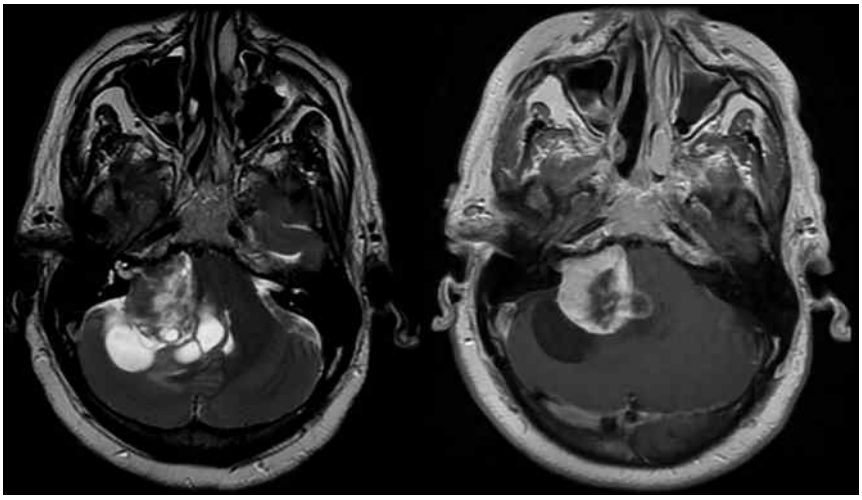


Fig. 19.11 Case 1. MRI at original presentation showed a 4-cm partially cystic tumor at the cerebellopontine angle consistent with vestibular schwannoma. Axial T2-weighted (*left*) and T1-weighted *(right)* postcontrast images showed the heterogeneous tumor with cystic components exerting mass effect on the cerebellum, brainstem, and fourth ventricle.

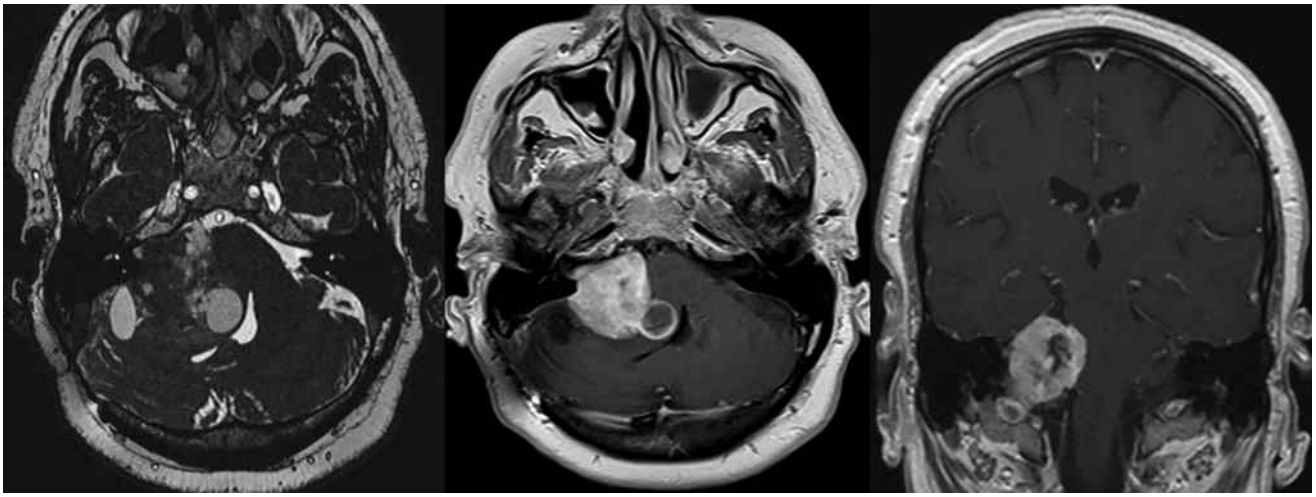


Fig. 19.12 Case 1 after first surgery. Interim MRI 6 months after subtotal resection via a retrosigmoid approach revealed progression of the known vestibular schwannoma residual. Axial T2-weighted constructive interference in steady state (*left*), axial (*center*), and coronal (*right*) T1-weighted postcontrast sequences showed enlargement of the enhancing, heterogeneous residual tumor with a cystic component projecting into the middle cerebellar peduncle.

House–Brackmann grade II facial nerve function on the right. He also had persistent gait instability and profound right upper extremity dysmetria.

Anatomical and Therapeutic Considerations

The patient presented with a progressive residual giant vestibular schwannoma and symptoms of both cranial nerve and brainstem compression. The tumor extended laterally in the IAC with widening of the porus acusticus, had a medial cystic component that projected into the fourth ventricle with significant compromise of the middle cerebellar peduncle, and extended superiorly to the trigeminal nerve, compressing and displacing it. The cystic components and bright, heterogeneous enhancement pattern suggested a soft consistency, which can be associated with increased vascularity and more difficult resection.

As discussed in the prior section, the utilization of a different approach from the prior surgery obviates the need to operate through a scarred surgical cavity. It also makes it far easier to answer the question, "What will you do differently this time?," since the prior retrosigmoid approach was less than adequate. Additionally, for this tumor, both the medial extension into the midline brainstem and the lateral extensions into a widened IAC would make complete resection via a repeat retrosigmoid approach more difficult. On the other hand, a presigmoid approach would provide the perfect surgical trajectory to the medial cyst, which is the portion of this tumor most threatening to the patient's brainstem function.

Given his younger age and rapid tumor growth, the goal of the surgery was to perform as aggressive a resection as possible, while trying to preserve facial nerve function. If, in balancing facial nerve preservation against complete resection, a small residual was left behind, then he would be a good candidate for radiosurgery for a small residual along the facial nerve. However, it was made clear to the patient before surgery that the goal of the operation was to be as complete a removal as possible, in order to prevent the possibility of multiple recurrences.

In order to maximize the advantages of access to both the "blown-out" IAC and especially to the most medial aspects of the tumor, we planned to expand our presigmoid translabyrinthine

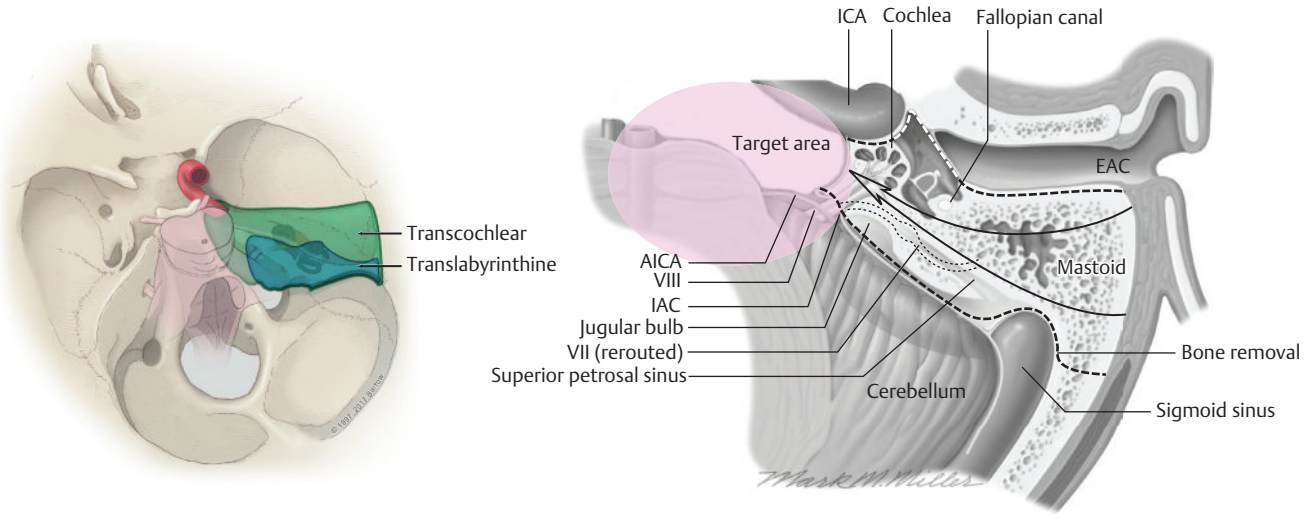


Fig. 19.13 The transcochlear approach. Compared to translabyrinthine approach, the transcochlear approach places the surgeon further anteriorly, making the line-of-sight more lateral-to-medial. This improves exposure of the midline structures and ventral brainstem. This exposure, however, requires the surgeon to work both anterior and posterior to the facial nerve, and in most cases, the nerve is transposed posteriorly, putting it at high risk for postoperative dysfunction. AICA, anterior inferior cerebellar artery; EAC, external auditory canal; IAC, internal acoustic canal; ICA, internal carotid artery; VII, facial nerve; VIII, vestibulocochlear nerve.

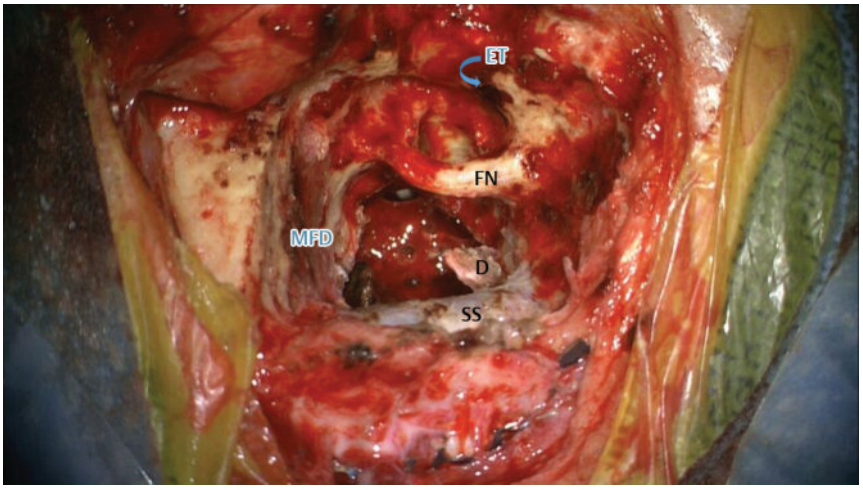


Fig. 19.14 Intraoperative image: transcochlear approach. In this patient, the transcochlear approach was made with the facial nerve (FN) left in its bony canal to preserve its function. The resection of the large right-sided vestibular schwannoma took place anterior and posterior to this arch-like structure, sometimes called the "fallopian bridge" because of its shape. D, posterior fossa dural flap; ET, eustachian tube; FN, facial nerve (in bony canal); MFD, middle fossa dura; SS, sigmoid sinus.

approach even further by utilizing a modified transcochlear approach (**Fig. 19.13**). We did not plan posterior transposition of the facial nerve, so as to maintain the possibility of an excellent facial nerve outcome, instead leaving the descending mastoid segment of the nerve protected in its bony canal with its vasculature intact.

Description of the Technique

The patient was positioned supine on the table with his head turned to the left and the ear folded forward. A C-shaped skin incision was made entirely anterior to the prior, traditional, linear retrosigmoid incision and a periosteal Palva flap was created. A wide mastoidectomy was performed, with the sigmoid sinus skeletonized from the lateral takeoff to the jugular bulb. The descending facial nerve was also skeletonized, but left in its canal with a thin shell of bone protecting it. A labyrinthectomy was performed after which the posterior, superior, and inferior walls of the IAC were identified, skeletonized, and removed. The blind sac of the ear was next created at this point. The external auditory canal skin was dissected free from the cartilage, everted, and sutured closed. The cochlea was then drilled away—the culmination of the bony removal was an approximately 300 degrees circumferential exposure of the tumor in the IAC with the facial nerve skeletonized from the IAC to the stylomastoid foramen, allowing access corridors to the tumor both anterior and posterior to the facial nerve (**Fig. 19.14**).

The presigmoid dura was opened in an H shape with superiorly and inferiorly based flaps; the tumor borders superiorly, inferiorly, and most posteriorly were easily identified; however, the border along the cerebellum was quite obscure because of scarring from the prior operation. The facial nerve probe was used to stimulate along the tumor borders to confirm the facial nerve was not in an aberrant position, and then the tumor was debulked. It appeared to be of superior vestibular nerve origin, as the vast majority was superior to the foramen of Luschka and most structures were displaced inferiorly. This was also confirmed at the fundus of the canal where the tumor was most adherent to the facial nerve. The facial nerve takeoff was then

identified on the brainstem, and it stimulated quite easily. The rest of the nerve was attenuated and significantly splayed by the tumor. Sharp dissection allowed separation of the tumor from the nerve, this time working laterally to medially, but it was significantly stuck at the porus.

The medial cyst was left for last, once the bulk of tumor was gone. The cyst wall could not be safely separated from the pia of the middle cerebellar peduncle, so the cyst itself was widely opened and marsupialized into the CPA cistern, leaving only a portion of the translucent cyst wall along the middle cerebellar peduncle. Ultimately, a near-total resection was completed with a very thin carpet of tumor remaining along the facial nerve. Motor evoked potential (MEP) response of the nerve was not significantly changed throughout the dissection, with stimulation intensity going from 130 V at the beginning to only 150 V at the end.

Aftercare

The patient's postoperative course was uncomplicated. MRI performed on postoperative day 2 showed the thin area of the intended residual (**Fig. 19.15**). His facial sensation improved and his facial nerve function improved to a House–Brackmann grade I. Clinically, the patient recovered well, save for a persistent pseudomeningocele treated with lumbar drainage and a subsequent wound reexploration 1 month postoperatively, which was uncomplicated and curative.

■ Case Presentation 2

A 45-year-old woman presented with a slow decline in right-sided hearing, vertigo, imbalance, and right tongue and chin numbness. Preoperatively her facial nerve function was normal and she maintained serviceable hearing on the right side. MRI revealed a 3.5-cm right vestibular schwannoma (**Fig. 19.16**).

Anatomical and Therapeutic Considerations

In addition to vestibulocochlear symptoms, the patient presented with progressive symptoms from the large tumor's mass

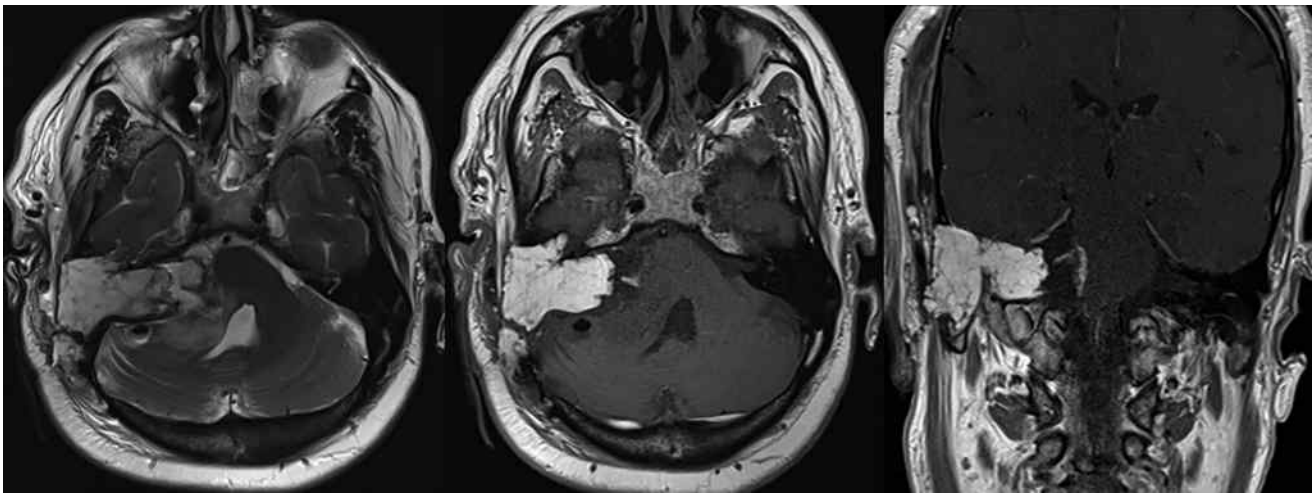


Fig. 19.15 Case 1 after transcochlear approach. MRI obtained on postoperative day 1 after transcochlear resection of the right-sided progressive residual cystic vestibular schwannoma with abdominal fat graft. The axial T2-weighted image (*left*) showed resolution of the mass effect on the middle cerebellar peduncle and fourth ventricle with persistent edema of the peduncle. Axial (*center*) and coronal (*right*) T1-weighted postcontrast images showed a tiny area of enhancement along the peduncle and at the origin of the facial nerve where the wall of the cyst could not safely be resected.

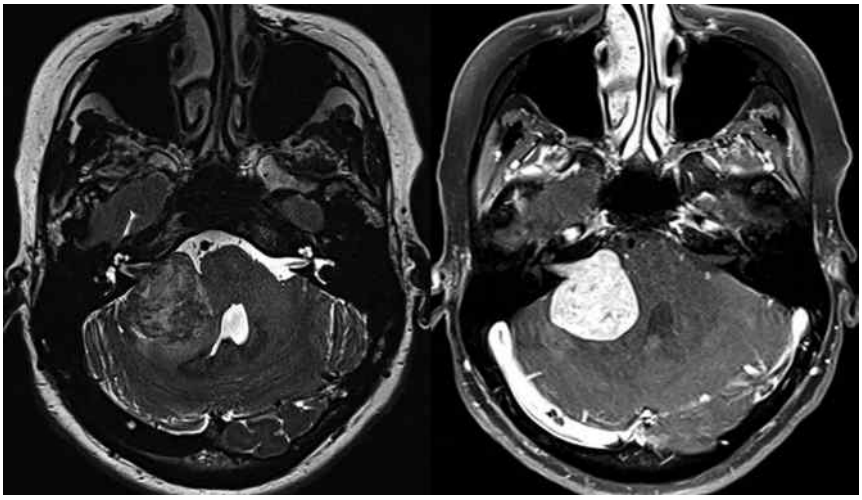


Fig. 19.16 Case 2. Preoperative axial T2-weighted constructive interference in steady state (*left*) and T1-weighted post-contrast (*right*) MRI revealed a 3.5-cm right vestibular schwannoma with mass effect on the cerebellar peduncle and fourth ventricle.

effect on the trigeminal nerve and brainstem. Though she did have serviceable hearing on an audiogram, she had switched her "telephone ear" from her right to the left. She was a young, otherwise healthy woman with normal facial function. Therefore, the primary goal of surgery was to remove enough of the tumor to provide long-term cure while preserving facial nerve function. On the preoperative MRI, the tumor itself was round and smooth, and from the exerted mass effect and somewhat decreased T2 signal, appeared likely to be somewhat firm. Firmness in a vestibular schwannoma can convey an advantage during resection, as the tumor will tend to push critical structures out of the way without adhering to them, making dissection simpler. Given the size of the tumor and low chance of preserving serviceable hearing regardless of approach, a translabyrinthine approach was recommended.

Description of the Technique

The same positioning, skin incision, and initial exposure as the transcochlear procedure described above were performed. After labyrinthectomy, the wall of the IAC was identified and drilled out, providing wide exposure. The jugular bulb was high and required drilling the bone medial and anterior to it. The dura was thick and opened from just anterior to the sigmoid sinus linearly down to the porus acusticus, with superior and inferior cuts creating an H-shape incision with superiorly and inferiorly based dural flaps. Prior opening of the cochlear aqueduct during exposure provided initial CSF drainage and opening the arachnoid in the cistern inferior to the tumor allowed further CSF drainage and relaxation of the cerebellum. The tumor was, as expected, firm with smooth contours.

After significantly debulking the tumor, the plane between the tumor and cerebellum was easily identified. The eighth nerve trunk was identified and its components were left in place initially, to serve as additional protection for the facial nerve. The facial nerve was then identified at the brainstem and stimulated at 0.1 mA with a brisk response. Working from medial to lateral this time, the tumor was dissected off the facial nerve up to the porus, where again it was very adherent; the facial nerve was pushed anterior to the lip of the porus and was significantly splayed. Working next in a lateral-to-medial fashion starting at the fundus medially, the superior and inferior vestibular nerves were cut and easily reflected off the facial nerve in the IAC.

The tumor was rolled out of the IAC until reaching the porus, where it was again stuck. Sharp dissection was used to separate the tumor from the facial nerve working from lateral to medial. A very thin portion of tumor was left on the facial nerve at the porus, and it was carefully coagulated in situ.

Aftercare

Postoperatively, the patient's right facial sensation improved to normal. She maintained normal House–Brackmann I facial nerve function. MRI done on postoperative day 2 showed no visible enhancing residual to the neuroradiologists. Her course was uncomplicated, and she was discharged home.

■ Case Presentation 3

A 51-year-old woman presented with 3 months of mild left-sided hearing loss and many years of intermittent vertigo. She had class A hearing on her audiogram. MRI showed a 2.7-cm left CPA tumor (**Fig. 19.17**).

Anatomical and Therapeutic Considerations

The patient's preoperative MRI showed a large, enhancing tumor that was almost entirely extracanalicular. The adjacent dura also enhanced, suggesting a small dural tail, and there was a question of hyperostosis on that side. These radiographic findings, combined with the patient's good hearing status, made meningioma a reasonable, if small, possibility.

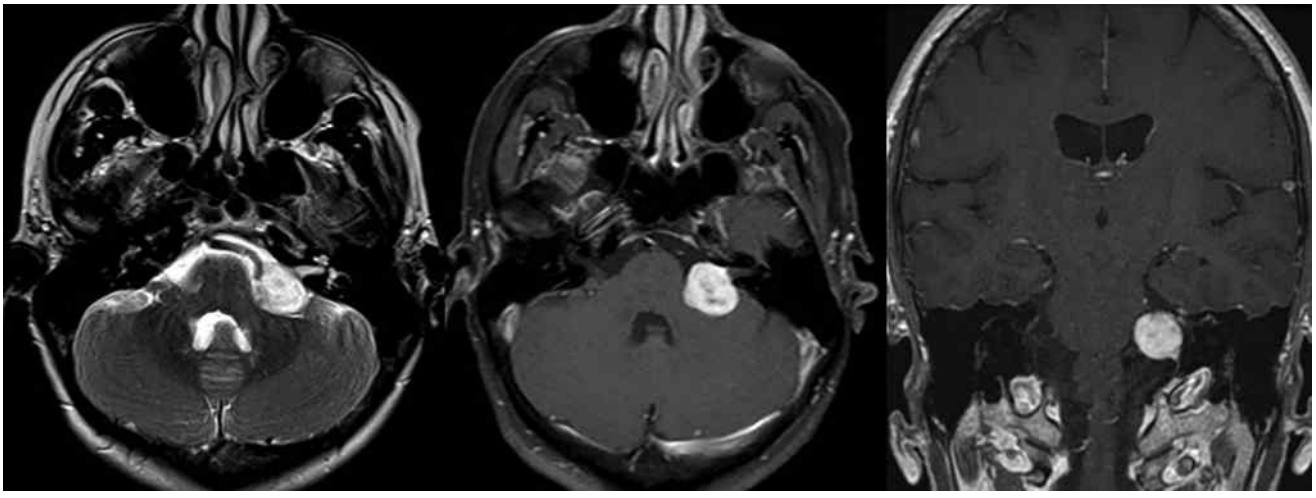


Fig. 19.17 Case 3. Preoperative MRI for a 51-year-old woman presenting with mild left-sided hearing loss and chronic intermittent vertigo. The axial T2-weighted image (*left*) showed a round, hyperintense tumor with mild mass effect on the peduncle. The axial T1-weighted postcontrast image (*center*) showed that the lesion was a maximum of 2.7 cm in diameter and was almost entirely extracanalicular. The coronal image (*right*) showed the slight adjacent dural enhancement at the inferior aspect, suggestive of a possible dural tail.

Given this possibility and her preserved hearing, surgery via a retrosigmoid approach to attempt hearing preservation was recommended.

Description of the Technique

The patient was positioned supine with the head placed on a foam donut and turned to the right. A gently curved, anteriorly based C-shaped incision was made and the skin elevated with a separate periosteal (Palva) flap. The neurotologist drilled away the bone overlying the sigmoid sinus from the transverse–sigmoid junction to the jugular bulb, exposing 2 mm of retrosigmoid dura. The dura was freed from the overlying retrosigmoid bone, and the footplate of the craniotome was then placed at the sigmoid–transverse junction in the epidural space. A 2.5-cm craniotomy was then easily turned with the craniotome. The dura was opened parallel to the sigmoid and transverse sinuses.

Under the operating microscope, CSF was released from the cisterns inferiorly. No retractors were used, and the tumor fell out of the way posteriorly. The tumor encountered was smooth, round, and soft, with one large nodule protruding posteriorly into the cerebellar hemisphere and middle cerebellar peduncle. It had the typical appearance of a vestibular schwannoma and seemed to be of superior vestibular origin, with only 5 mm of extension into the IAC. The majority of the tumor was debulked until the walls were very thin and could be folded in. It was noted to be positioned on the posterior face of the trunk of the eighth nerve. From this retrosigmoid exposure, the facial nerve dissection off the tumor capsule proceeded in the only available direction, medial to lateral. This dissection halted at the porus, where the tumor became adherent and where there was insufficient exposure to identify the proper dissection plane anterior to the tumor.

The neurotologist now drilled off the posterior wall of the IAC to a depth of approximately 6 mm. The facial nerve was displaced superiorly. The cochlear nerve was very attenuated and, although anatomically preserved, the auditory brainstem response became unreliable and ultimately lost as the final portion of tumor was removed at the porus. A GTR was achieved. The facial nerve stimulated at the brainstem at 0.05 mA at the end of the dissection, and the facial MEP was only minimally changed from 130 to 135 V. In closing, the dura was tightly reapproximated and the patient's own bone was plated back into position. Any exposed gaps and the sigmoid sinus were covered by a harvested, abdominal fat autograft.

Aftercare

The postoperative course was uncomplicated and the patient had normal facial nerve function. She did, however, lose hearing in the left ear and was fitted with a BiCros hearing aid as an outpatient. She had unusual headaches that gradually subsided over the subsequent months.

■ Commentary

Many factors contribute to surgical planning for large vestibular schwannomas, and the cases above highlight some factors that are unique to choosing an approach for these tumors. In general, for recurrent tumors it is preferable to choose an approach that is different from the initial approach to avoid dissection along a scarred corridor. This may be aided by reviewing the patient's original preoperative imaging or operative report to identify the limitations of the initial approach, particularly if the first resection was an unplanned subtotal resection or resulted in unexpected complications. Thus, when faced with a large recurrent tumor after a retrosigmoid approach, as in Case 1, we would choose to perform a translabyrinthine or transcochlear approach for the second resection.

Of the three primary approaches for vestibular schwannoma resection, the retrosigmoid or translabyrinthine approaches are most appropriate for large tumors. The retrosigmoid approach provides a wide view of the posterior fossa contents from the tentorium to the foramen magnum and is frequently used by neurosurgeons for a variety of pathologies, making it the most commonly utilized approach at many centers. Compared to the translabyrinthine approach, the obvious, and we would claim only, benefit of the retrosigmoid approach is the possibility of hearing preservation. In meta-analyses, the literature suggests that for large tumors, there may be lower rates of facial nerve dysfunction, higher rates of headache or CSF leak, and equivalent rates of residual tumor and complication with the retrosigmoid approach when compared to the translabyrinthine approach.

Our experience suggests the opposite of what is seen in large retrosigmoid series, possibly because of the close collaboration between neurosurgery and neurotology. It is particularly hard to understand why otherwise firm supporters of the tenets of skull base surgery—bone removal rather than brain retraction, keyhole exposures, decreasing the depth of the field—forget that it is the translabyrinthine approach that adheres to all of these principles and not the retrosigmoid approach. Many neurosurgeons still use retractors to keep the cerebellum out of their view in a retrosigmoid approach. In contrast, when performing a translabyrinthine approach, the exposure is achieved through bony drilling and does not require use of retractors. Furthermore, it is the translabyrinthine approach, not the retrosigmoid approach that makes the field shallower for the surgeon, improving the surgeon's control, dexterity, and reach to the midline. Again, whereas the literature might indicate otherwise, we find that the ability to dissect the facial nerve from tumor in both directions, lateral-to-medial and medial-to-lateral, represents another advantage of the translabyrinthine approach.

Consistent with the literature, we have noted higher incidence of postoperative headache with the retrosigmoid approach, and a greater chance of leaving residual tumor in the lateral IAC. This lateral residual is a particular problem for young patients, opening them to the risk of multiple recurrences and surgeries with ever-increasing risk over a long lifetime. For these reasons, at our institution the translabyrinthine approach is most frequently chosen for resection of large tumors; if the patient in Case 1 had presented to our institution after initial diagnosis, he likely would have undergone a translabyrinthine approach for his first surgery. One criticism of the translabyrinthine approach is the large amount of time it takes to remove the petrous temporal bone adequately and precisely. However, by refining the collaboration between neurosurgery and otolaryngology, we have maximized efficiency and minimized

operative time on translabyrinthine cases, making it the preferred approach even for less optimal surgical candidates. In older, less healthy patients, we prefer a "quick" translabyrinthine approach, because it is a smaller opening, exposes less brain, and provides a better trajectory to large tumors, especially at their medial, hardest-to-reach portion.

There is clearly room for debate when a patient presents with a large tumor but has retained serviceable hearing. However, the rates of preserving hearing in patients with large tumors are very low (<10%), regardless of preoperative hearing status. If, during attempted hearing preservation, fibers of the vestibular nerve are left in place and the patient still loses hearing, there is also a higher risk of persistent vestibular symptoms postoperatively. Thus, for most patients presenting with large tumors, even in the context of serviceable hearing, as in Case 2, the translabyrinthine typically remains our approach of choice.

The patient described in Case 3 highlights the factors that favor a retrosigmoid approach. She had normal hearing on her preoperative audiogram; her tumor was smaller than the other two examples, and was predominantly extracanalicular. The odds of hearing preservation were more favorable. Tumors that follow the "rule of roundness," with smooth, round borders and a T2 hypointense core, tend to be firm and push the nerves out of the way, improving the chance of hearing preservation. Similarly, tumors that do not invaginate into or involve the foramen of Luschka also may have higher rates of hearing preservation, since the vestibulocochlear nerve is free of the tumor over its medial extent. Clues of this can be gleaned from the MRI scan from the extent of compression of the flocculus and the choroid plexus of the foramen of Luschka. If these are free, then the vestibulocochlear nerve root, which enters the brainstem in close proximity, is likely as well.

In addition, the details provided in modern MRI sequences often allow prediction of the nerve of origin; tumors growing superiorly that appear to be of superior vestibular nerve origin are preferable for a hearing preservation approach. A preoperative videonystagmography, cold and warm caloric testing, might be useful as these evaluations examine superior vestibular nerve function. Tumors that do not extend anteriorly past the lip of the IAC are also possible candidates for hearing preservation, since the cochlear and facial nerves will not be deformed anterior to the porus. Furthermore, a cap of CSF seen on the constructive interference in steady state or fast imaging employing steady-state acquisition sequence, between the lateral end of the tumor and the habenula perforata of the cochlea, also portends to better chances of hearing preservation. The ability to identify these important details on imaging in the context of a patient's clinical presentation allows for selection of the most reasonable candidates for resection of large vestibular schwannomas via the retrosigmoid approach for attempted hearing preservation.

In the final analysis, however, we suspect that the single most important factor in achieving superior results for large vestibular schwannomas is the close collaboration between neurosurgeon and neurotologist. Within this partnership, the choice of approach is made through discussion and debate, and throughout what might be difficult operations, two experienced surgeons with different perspectives can encourage, correct, and guide each other, to achieve the best possible outcome for the patient.

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20 Jugular Foramen (Intracranial)

Ken Matsushima and Michihiro Kohno

Keywords: jugular foramen, suprajugular approach, endoscopic, retrosigmoid keyhole approach

■ Case Presentation

A 39-year-old, otherwise healthy man presented with vertigo, tinnitus, and hearing disturbance. A head computed tomography (CT) scan showed a left cerebellopontine angle tumor, and the patient was referred to our hospital.

Questions

1. What is the differential diagnosis of a mass that extends into the jugular foramen?
2. How does the differential diagnosis influence the next step in diagnosis and management?
3. What is the most common presenting symptom for jugular foramen neuromas, and does it influence surgical strategy?

■ Diagnosis and Assessment

The magnetic resonance imaging (MRI) showed a round, enhancing cerebellopontine angle tumor with a cystic component, extending into the upper jugular foramen (**Fig. 20.1**). The jugular bulb was not invaded by the tumor, and the patency of the sigmoid sinus and the internal jugular vein was confirmed by digital subtraction angiography (DSA). The bone window on CT revealed a smoothly eroded and enlarged glossopharyngeal meatus. There was no intense tumor stain on DSA.

The differential diagnosis for this tumor includes schwannomas, meningiomas, and paragangliomas (glomus jugular tumors). We routinely perform contrast CT scans, enhanced MRIs, and DSA preoperatively to differentiate and evaluate these tumors. MRIs include enhanced T1-weighted sequences, pre- and postenhanced heavily T2-weighted MR cisternography (constructive interference in steady state/fast imaging employing steady-state acquisition), diffusion-weighted imaging, and susceptibility-weighted imaging. The axial and coronal sections in heavily T2-weighted MR cisternography allow detailed evaluation of surrounding neurovascular structures. Diagnostic features suggestive of schwannoma include cystic components, osseous erosion of the foramen, and spotty signal voids inside the tumor on susceptibility-weighted imaging. In comparison, meningiomas are more frequently associated with significant tumor "blush" on DSA, a distinctive dural tail sign of the tumor surface, calcification, and hyperostosis, without erosion of the foramen. Paragangliomas typically show aggressive destruction and invasion of the surrounding structures, including the jugular foramen and bulb, presence of flow voids within the mass, and strong tumor stain with early arteriovenous shunting on DSA.

In our experience, these radiological assessments can distinguish schwannoma from other tumors in almost every case. The findings in our patient included a cystic component, bony erosion, minimal tumor blush, and extension into the jugular foramen. Importantly, there was also a lack of involvement of the internal acoustic meatus. The most likely diagnosis was therefore a jugular foramen schwannoma. As to which one of the lower cranial nerves gave rise to the tumor, current diagnostic technology is still insufficient to provide us the answer routinely.

Since total resection of these jugular foramen tumors can be potentially curative, the avoidance of approach- and resection-related neurological morbidities is critical for management of these benign tumors. The multidisciplinary evaluations by otolaryngologists and speech therapists, including audiometric tests, swallowing, and speech tests, should be performed before and after surgery for these patients, even if these symptoms seem insignificant. In cases where the favored diagnosis before surgery is meningioma or paraganglioma, one should consider preoperative embolization of the feeding arteries. For paragangliomas, assessment for catecholamine secretion should also be performed.

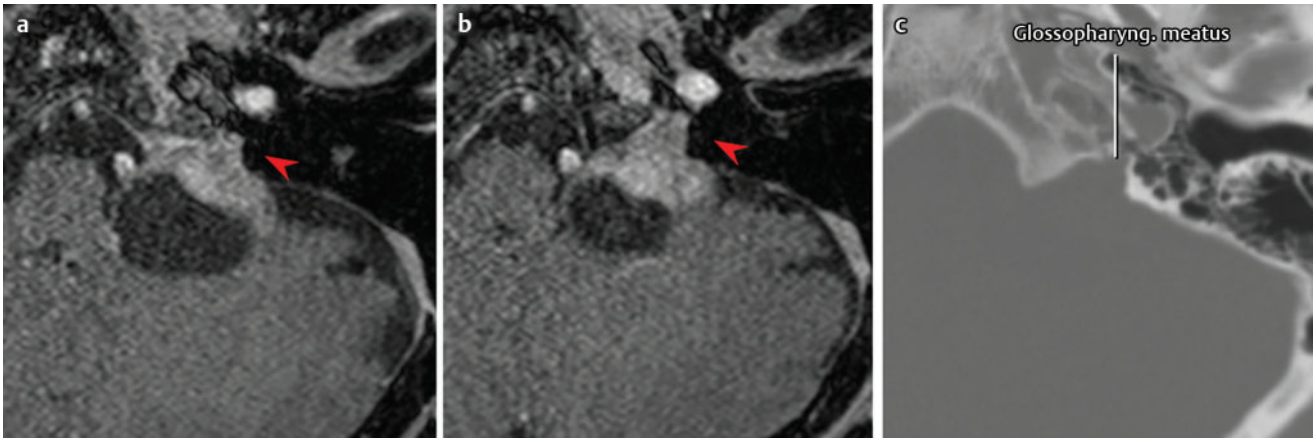


Fig. 20.1 Preoperative radiological images. (a, b) The enhanced T1-weighted MRIs showed a round enhancing cerebellopontine angle tumor with cystic component, extending into the upper jugular foramen (*red arrowheads*). **(c)** The bone CT scan revealed a smoothly enlarged glossopharyngeal meatus.

■ Anatomical and Therapeutic Considerations

The jugular foramen is located at the posteroinferior aspect of the petro-occipital fissure. From an intracranial point of view, its shape resembles a pear, with a narrow anteromedial part and a larger posterolateral part. Classical teachings divide the jugular foramen also into two anatomical portions: the anteromedial pars nervosa, traversed by cranial nerve IX and the inferior petrosal sinus (IPS), and the posterolateral pars venosa, traversed by cranial nerves X, XI and the jugular bulb (**Fig. 20.2**). More recent data suggest that there may in fact be three compartments, the petrosal part with the IPS, the sigmoid part with the sigmoid sinus, and an intrajugular part between the two venous parts, through which the cranial nerves travel.

Options for Approach

Regardless of the compartment scheme one uses, the jugular foramen is one of the most difficult regions to access surgically because of its deep location, the presence of surrounding vital structures, and the frequent intra- and extracranial extension of the lesions through the foramen. Jugular foramen schwannomas have been classified into three types: intradural, dumbbell-shaped, and triple dumbbell-shaped tumors. The surgical approach should be selected based on tumor type and anatomical extensions. In general, approaches that require sacrifice of hearing should be avoided. Auditory symptoms are quite common for jugular foramen schwannoma, and this symptom frequently improves after surgery. In our experience with these patients, almost 40% of the patients had hearing disturbance before the surgery, and half of them improved afterward. Therefore, even if the preoperative hearing is unserviceable, the choice of surgical approach must preserve hearing.

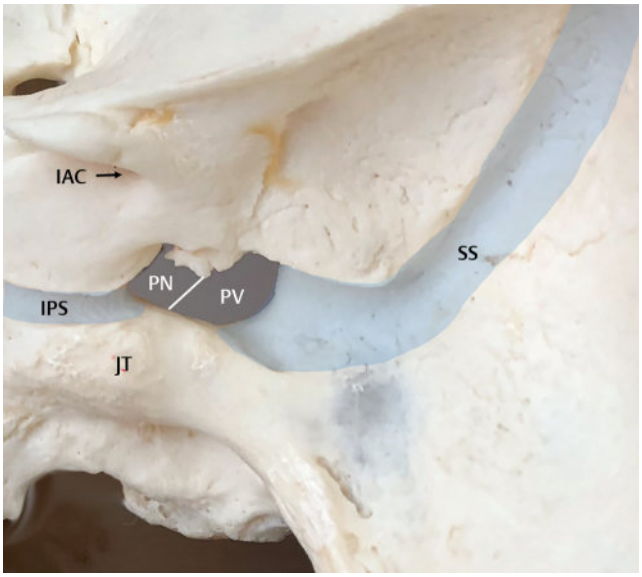


Fig. 20.2 Dry skull image of the right jugular foramen. The jugular foramen is located at the posteroinferior aspect of the petro-occipital fissure. It has a narrow anteromedial part and a larger posterolateral part. Classically described, the anteromedial pars nervosa (PN) contains the cranial nerve IX and the inferior petrosal sinus (IPS), and the posterolateral pars venosa (PV) contains the cranial nerves X, XI and the jugular bulb. Most recent studies divide the foramen into three compartments (see text). IAC, internal acoustic canal; JT, jugular tubercle; SS, sigmoid sinus.

Skull base approaches to the jugular foramen go by many names, but most of them involve extradural drilling to open the jugular foramen from either a posterolateral or straight lateral trajectory. Since they were designed for pathology that either invade or involve the jugular bulb, the sigmoid sinus and/or jugular vein are ligated in these approaches. In our series of jugular foramen schwannomas, half of the tumors had significant intraosseous and extracranial extensions, and these required the transjugular (trans-sigmoid) approach for total removal.

However, for the patient presented here, the major portion of the schwannoma was intracranial. Although there is certainly tumor inside the foramen, there was little, if any, extracranial extension of the tumor. The jugular bulb was patent, and there was no tumor invasion into it. For these reasons, large skull base approaches, such as the transmastoid–transjugular, or postauricular, transtemporal approaches would be counterproductive. The far-lateral supracondylar approach is perhaps worthy of consideration. A variant of the transcondylar approach, the drilling of the occipital condyle in this approach is focused more superiorly, as the name implies. The bone is removed toward the roof of the hypoglossal canal and the drilling eventually reaches the jugular tubercle. Although it "opens" the jugular foramen for more surgical freedom, the drilling around the jugular bulb is technically challenging in this approach. Extensive condylar removal can also put structural stability at risk at the craniocervical junction.

When the main bulk of the tumor is located intradurally, our approach of choice is the retrosigmoid approach. However, if a tumor extends into the upper part of the jugular foramen, as in our patient, the intraforaminal part may be difficult to remove solely by the conventional retrosigmoid approach. The clinical dilemma is whereas a radical removal of the intraforaminal portion requires extensive skull base approaches, leaving this portion behind simply allows it to grow back.

Approach of Choice

To solve this problem, the retrosigmoid suprajugular (or infralabyrinthine) approach has recently been developed. It allows access not only to the cisternal portion but also intraosseous extensions of the tumor, without sacrificing the sigmoid–jugular venous drainage system. Compared to more extensive approaches, the avoidance of extensive bone removal in this approach may reduce complications. The intradural suprajugular drilling enables access and removal of intraforaminal tumors, just as transmeatal drilling allows the removal of the intracanalicular part of vestibular schwannomas, or suprameatal drilling for access to the Meckel cave. In our experience of more than 60 surgeries of jugular foramen schwannomas, the retrosigmoid approach was selected in more than 40% of the cases, and suprajugular drilling was done in approximately half of them.

After careful consideration of all the clinical data for our patient, the retrosigmoid suprajugular approach was chosen with the aim of total tumor removal.

Questions

1. What kind of monitoring is essential for this operation?
2. What are the potential complications of this approach and how would you manage the endotracheal tube after surgery?

Description of the Technique

Before surgery, we routinely check the dominancy of the sigmoid sinus, the height of the jugular bulb, and its anatomical relationship with the tumor. The location and extent of the air cells, and the surrounding vascular structures including the posterior inferior cerebellar artery and superior petrosal vein, were also carefully noted. In most cases of intradural jugular foramen schwannoma, the tumor compresses lower cranial nerves caudally and cranial nerves VII and VIII cranially. If the tumor compresses the jugular bulb significantly, other collateral drainage routes including emissary veins may develop which may affect the surgical approach and tumor dissection (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: posterolateral

Craniotomy: retrosigmoid

Modifier: removal of the roof of the jugular foramen

Because of the concern for dysphagia and vocal cord paralysis, intraoperative neuromonitoring is indispensable. We routinely perform electromyograms of cranial nerves V, VII, and lower cranial nerves, auditory brainstem response, and somatosensory evoked potentials during this surgical approach. Continuous monitoring of evoked cranial nerve X electromyograms, in addition to free running spontaneous cranial nerve X electromyography and its occasional stimulation, is essential in avoiding nerve damage during the dissection (**Fig. 20.3**). Furthermore, for large tumor that significantly displaces the brainstem, motor evoked potentials should be monitored additionally.

In a park bench position with neck flexed, a C-shaped retroauricular incision was made. For a lateral suboccipital retrosigmoid craniotomy, an ultrasonic Doppler was used to confirm that the edges of the transverse and sigmoid sinuses and their junction were exposed. The posterior part of the sigmoid sinus was exposed in order to obtain an adequate operative field and lighting (**Fig. 20.4a**). Opening the foramen magnum should be considered if the tumor were large. After making a U-shaped dural incision and reflecting dura laterally over the sigmoid sinus, the cerebrospinal fluid (CSF) was released sufficiently from the cerebellomedullary cistern to relax the cerebellum. This enabled its gentle retraction (see **Operative Setup**).

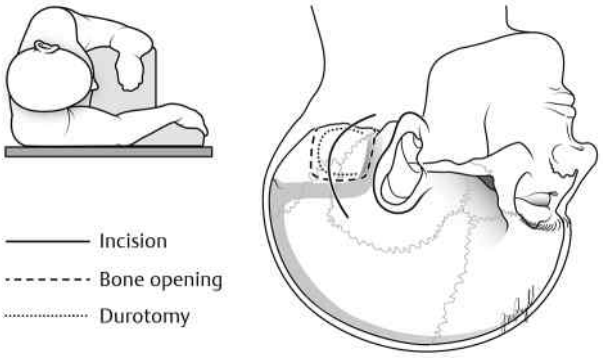
Operative Setup

Position: lateral, park bench

Incision: retroauricular, C-shaped

Bone Opening: lateral suboccipital 4 cm^2

Durotomy: U-shaped, based on the sigmoid sinus



Monitoring and Its Impact on Resection

The cranial nerve XI was identified in the cistern, and the cranial nerves IX and X were dissected from the caudal part of the tumor. After identifying the junction of cranial nerve X with the brainstem using stimulation of cranial nerve X electromyography, a ball-type monopolar stimulating electrode was placed on the proximal site of the cranial nerve X. In conjunction with a recording monopolar needle which was placed on the soft palate before surgery, the evoked cranial nerve X electromyogram was monitored with electrode 1-Hz stimulation during the intradural procedure (**Fig. 20.4c**). The supramaximal voltage (M-max) was recorded. Subsequently, continuous monitoring was started with the lowest electrical stimulation to evoke stable compound muscle action potentials. With this monitoring, we were able to monitor the function of cranial nerve X as we performed the tumor dissection, and this helps us with decision making

Fig. 20. 3 Setup for intraoperative neuromonitoring. The needle electrodes were placed on the frontalis, orbicularis oculi, and orbicularis oris muscles for cranial nerve VII electromyogram, as well as on soft palate for cranial nerve X electromyogram, sternocleidomastoid muscle, and tongue.

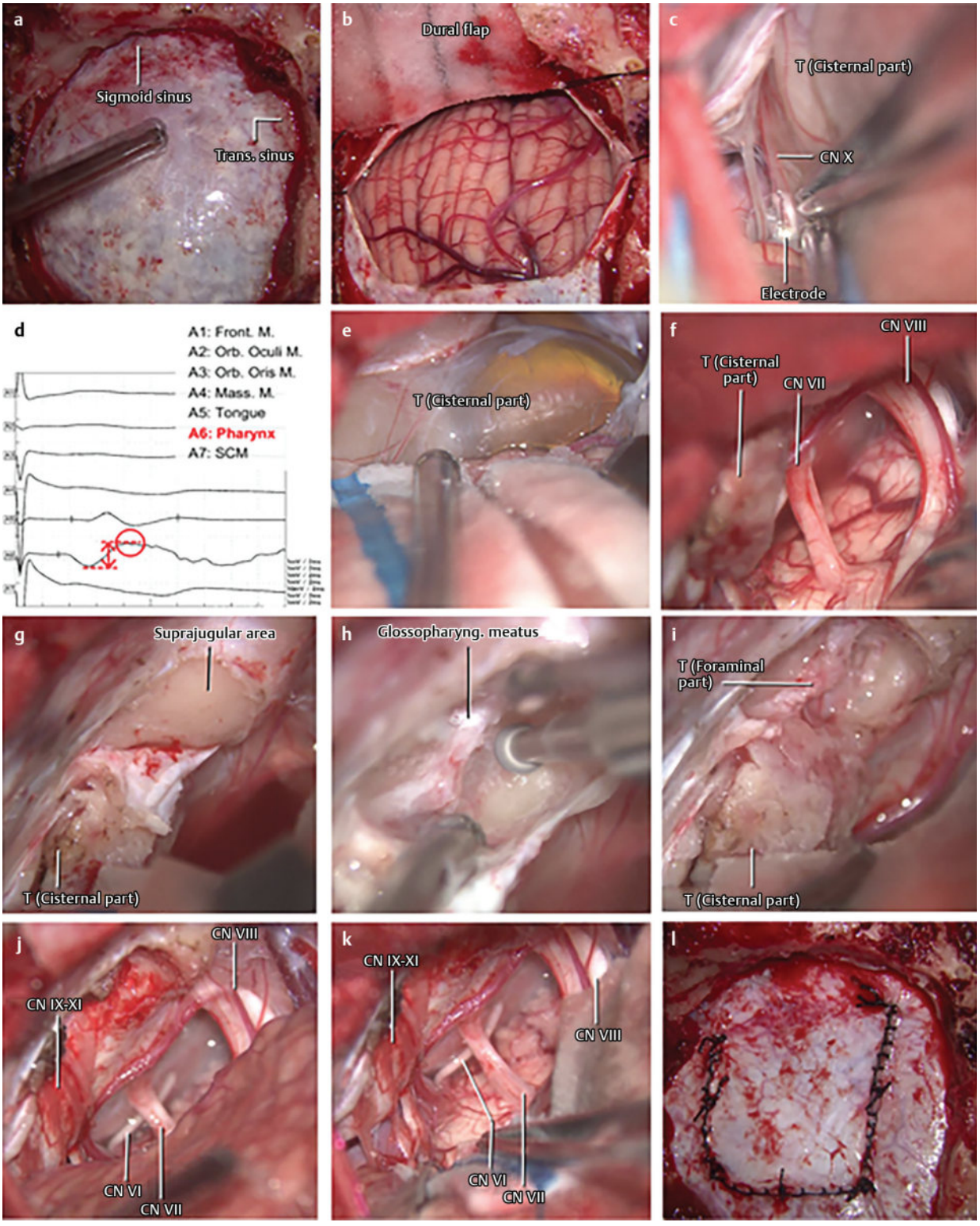


Fig. 20.4 Intraoperative images. (a) The posterior part of the sigmoid sinus was routinely exposed in order to obtain a sufficient operative field and lighting. **(b)** Incised dura was reflected over the sigmoid sinus and covered to prevent it from becoming dry and shrinking. **(c)** A ball-type monopolar stimulating electrode was placed on the proximal site of the cranial nerve X for its continuous monitoring. **(d)** The amplitude and latency of the evoked cranial nerve X electromyogram were monitored with 1-Hz stimulation during the intradural procedure. **(e)** Tumor with cystic component was exposed in the cerebellopontine angle. **(f)** After cystic puncture and resecting the cisternal tumor, the remaining tumor was observed at the intradural porus of the jugular foramen. **(g)** The dura of the suprajugular region was incised and dissected inferiorly. **(h)** The suprajugular drilling opened the glossopharyngeal meatus. **(i)** After opening the dura of the glossopharyngeal meatus, the foraminal part of the tumor connected with the remaining cisternal part of the tumor was identified. **(j, k)** After tumor removal, residual tumor was identified neither in the jugular foramen nor in the cerebellopontine angle. **(l)** The dura was closed in a watertight manner without graft materials under microscope.

during the procedure. Preservation of M-max of more than 30% of the control is the goal to avoid postoperative cranial nerve X functional deficits. As long as this was preserved, the resection continued until the tumor was completely removed. If, on the other hand, a drop in M-max were to be detected during the procedure, we might consider leaving some portions of the tumor behind, in order to preserve vagus nerve function.

Suprajugular Drilling

Piecemeal debulking of the cerebellopontine angle tumor and drainage of the cyst were performed after direct stimulation on the dorsal tumor surface to confirm that there were no motor nerve filaments. The cranial nerves VII and VIII were dissected meticulously from the cranial part of the tumor, while respecting the arachnoid planes. After removal of the cisternal component, the dura above the jugular foramen was incised and dissected inferiorly from the posterocaudal part of the internal acoustic meatus. As the dura is strongly adherent along the crease, the endolymphatic depression (endolymphatic fovea) was a good landmark for the posterolateral limit of the dural incision. The exposed roof of the jugular foramen was carefully drilled with a 2- or 3-mm high-speed diamond drill, with keen awareness of the location of the nearby lower cranial nerves and jugular bulb (**Fig. 20.4h**). The amount of drilling depended on the anatomical variation, pattern of tumor extension, and the degree of bone erosion. As schwannomas originating from the lower cranial nerves frequently occupy the ventral portion of the jugular foramen while displacing the jugular bulb dorsally, drilling around the glossopharyngeal meatus (the ventral part of the roof) is often required. Drilling the area deep to the endolymphatic depression (the dorsal part of the roof) may provide wider access inside the jugular foramen, but this puts the jugular bulb and sigmoid sinus, with their many anatomical variations, at risk. Fortunately, this is rarely needed.

The jugular bulb was encountered and skeletonized in our case. This method of drilling provided a route into the superior part of the jugular foramen and enabled removal of tumor extensions into the foramen. The intraoperative navigation system or angled-endoscopic assistance can increase the safety and efficacy of the foraminal drilling, which in turn, increases the likelihood of complete tumor removal. In our case, the intraforaminal tumor was dissected carefully from the lower cranial nerves and gross total tumor removal was achieved without M-max drop. Bone wax and fibrin glue were used to cover the bone defect completely to avoid postoperative CSF leakage. Lastly, the dura was closed in a watertight manner under microscope.

Surgical Pearls

1. Appropriate selection of the surgical approach based on detailed preoperative radiological evaluation and anatomical knowledge is critical for jugular foramen schwannoma surgery. Although hearing disturbance is one of the most common presenting symptoms, surgical approaches that sacrifice or endanger hearing function should be avoided since there is always a chance for hearing improvement.
2. The surgical goal for this benign tumor is maximizing tumor removal without any postoperative neurological deficits. Postoperative swallowing difficulty or vocal cord paralysis significantly affects the patient's quality of life. Intraoperative neuromonitoring including the continuous monitoring of evoked cranial nerve X electromyograms should be utilized for decision making on surgical strategy during tumor resection. Also, pre- and postoperative evaluations by otolaryngologists and speech therapists should be routine, even if the patient does not seem to have dysphagia or hoarseness.
3. It is crucial to carefully carry on each step of the procedures for skull base surgery. The sequential completion of the following steps is required to perform this approach: (1) internal decompression, (2) identification and dissection of the nerves around the tumor, (3) total removal of the cisternal part of the tumor, (4) the suprajugular drilling, and (5) removal of the foraminal part of the tumor.

■ Aftercare

Extubation was performed immediately after surgery and fiberoptic laryngoscopy did not reveal any vocal cord dysfunction. The laryngoscopy should be performed before restarting oral feeding. Postoperative MRI and CT scans confirmed complete resection of the lesion and showed the extent of osseous drilling (**Fig. 20.5a–c**).

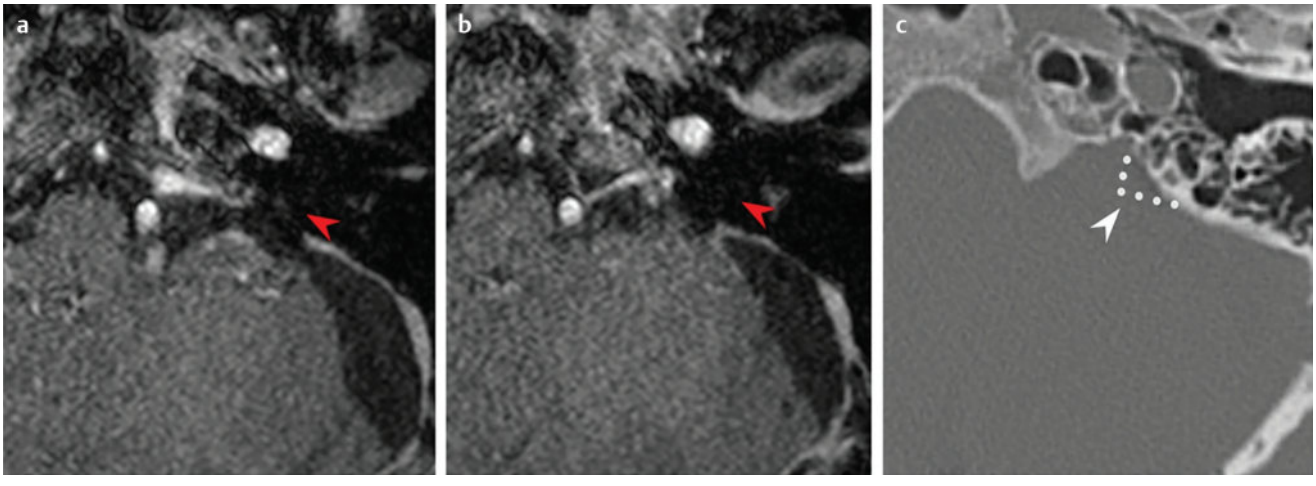


Fig. 20.5 Postoperative radiological images. (a, b) MRI confirmed absence of residual tumor (*red arrowheads*) and **(c)** CT scans showed the extent of osseous drilling around the glossopharyngeal meatus (*white arrowhead*).

The preoperative hearing disturbance improved after surgery (class D [PTA: 48.8 dB, SDS: 0%] to A [PTA: 13.8 dB, SDS: 94%] according to American Academy of Otolaryngology–Head and Neck Surgery [AAO-HNS] hearing classification) (**Fig. 20.6**). The patient has no neurological deficits or recurrence of tumor during 30 months of follow-up after operation.

■ Possible Complications and Associated Management

The drilling around the jugular foramen must be performed with utmost care. The meticulous consideration of the surrounding anatomy has already been emphasized, but with so many vital structures nearby, the surgeon must be aware of his or her environment while the drill is engaged. Copious irrigation will help prevent thermal injury to the nerves, and minimize the caustic effects of bone dust.

Careful swallowing evaluation must be done before allowing the patient to return to eating solid food. Despite the aforementioned laryngoscopy, and intraoperative monitoring of the lower cranial nerves, subtle swallowing deficits can lead to aspiration, which can completely derail the patients' recovery. The patients should also be monitored for CSF leak postoperatively, as this can occur not only through the incision, but also through the air cells around mastoid and jugular foramen.

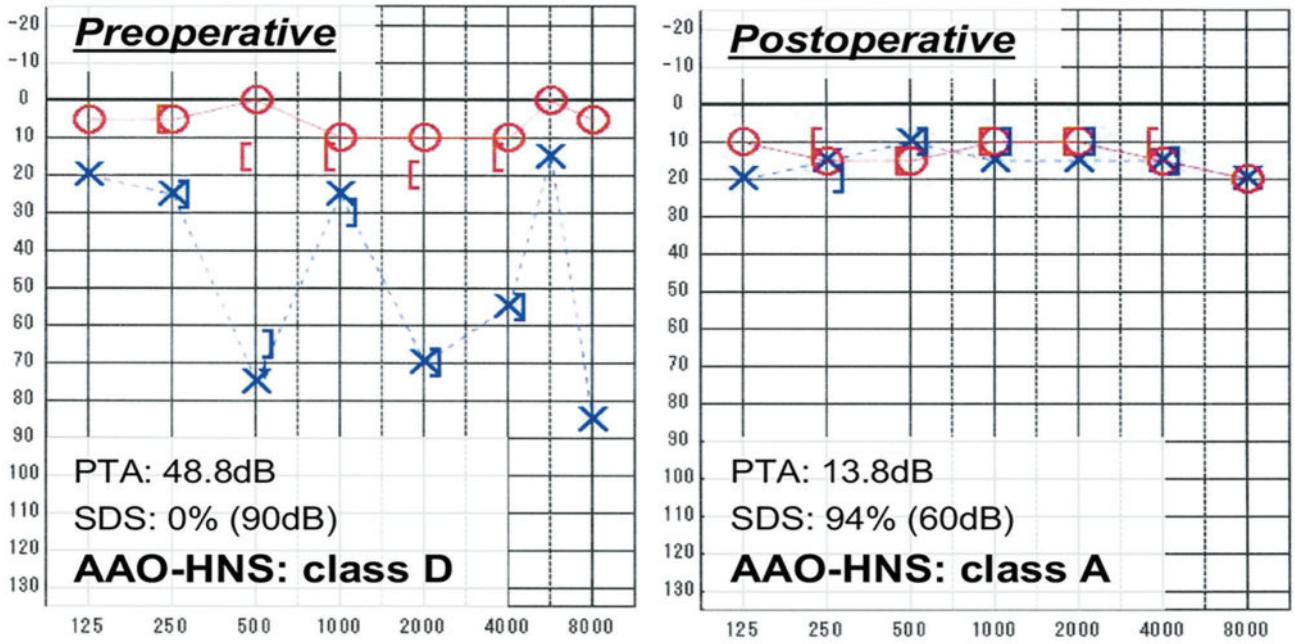


Fig. 20.6 Pre- and postoperative audiograms. Hearing disturbance significantly improved after surgery.

Perspective

Walter C. Jean

■ Introduction

When one examines the literature written on jugular foramen tumors in the recent decades, one sees a trend: the described operations became huge and monstrously complex, before returning to a more compact, simplified form. This "pendulum" swing from small-and-simple to large-and-complex and back again is seen repeatedly in other realms of skull base surgery, and there are two main driving forces behind it. The first represents the process of "learning." Surgeons, as they learn more about anatomy, started by being increasingly aggressive with bone removal and going through more anatomical structures. The operations take on more and more "trans-" terms, such as "transcondylar" or "transjugular," as this process evolves. The backswing of the pendulum starts as we recognize the morbidity associated with these same trans- terms, and the learning process takes us back to more compact and rational operations. The second driving force is technological advancement, and nowhere is the influence of this more evident than in the nearly ubiquitous use of endoscopy in skull base surgery. With enhancements in visualization of skull base pathology, new operations tend to be less invasive and smaller in scale.

While jugular foramen surgery went through this evolution, nomenclature followed the operations as they grew more complex. It was hard enough to understand the anatomy and surgical steps, but it became nearly impossible as surgeons called similar operations by very different names. However, whether they were named after famous surgeons, such as the various Fisch type operations, or recognized by acronyms, such as E.L.I.T.E (standing for "extreme lateral inferior transcondylar–transtubercular exposure"), these so-called skull base approaches all involved opening the jugular foramen extradurally, ligation of the jugular vein and sigmoid sinus, and sometimes, mobilization of the facial nerve as well.

Whereas there is still a proper application of these large transjugular approaches for tumors which have sizable extradural components (e.g., Fukushima's type B and C tumors), the conundrum is highlighted by a schwannoma that is mainly intradural but has a sizable foraminal component. While opening the foramen and eliminating the jugular vein seem to be "overkill," a simple retrosigmoid approach seems insufficient. Although nuances of the clinical issues were well-described in the previous section, we took a somewhat different approach.

■ Case Presentation

A 39-year-old man presented with a 6-week history of pulsatile tinnitus on the right. He also complained of difficulty swallowing solid foods and a mild headache. Physical examination was unable to detect any significant neurological deficits, and an office endoscopy showed normal vocal cord function on both sides. An MRI was performed, and this showed a jugular foramen tumor (**Fig. 20.7**). He was referred to our unit for further care.

■ Anatomical and Therapeutic Considerations

Once again, we have a tumor consistent with a jugular foramen schwannoma, and like the patient in the previous section, this patient sought medical care mainly because of hearing-related issues. The MRI showed a tumor that distorted the brainstem, and posited mainly in the intradural compartment. However, it also had a sizable foraminal component which must be addressed if we were to minimize the chances of tumor recurrence in this young healthy man.

Before arriving at a surgical solution, our surgical team made two assessments at the outset. First, we acknowledged that a lateral or posterolateral approach, with elimination of the jugular vein and sigmoid sinus, would run against the patient's best interest. This approach would be too "big" and there were better ways to eliminate the foraminal portion. Second, it was clear to us that a retrosigmoid approach was not enough, and the foraminal portion would be perpendicular to the line-of-sight of that approach and hidden from view.

Partially opening the foramen with the drill is one way to improve visualization of the tumor in the foramen. Drilling can be part of a far-lateral, transcondylar approach, and once again highlighting the complexity with nomenclature of this topic, a far-lateral approach to the jugular foramen would in reality involve drilling *above* the occipital condyle extradurally ("supracondylar") toward the jugular tubercle ("transtubercular") to partially open the foramen from below. The enlargement of the foramen would be at the inferior lip but most likely, the foraminal part of the tumor is near the superior part of the foramen.

As such, a better alternative may be the intradural, suprajugular opening that was described previously. A subtle variant of the technique has been described as "infralabyrinthine," which highlights the existence of the bony labyrinth nearby. In actuality, it is the endolymphatic sac that is most likely in harm's way, and the drilling superoposteriorly must be limited for its preservation (**Fig. 20.8**).

We concluded that preparation must be made for microscope-guided drilling above the foramen, whether one chose the term of "suprajugular" or "infralabyrinthine." However, we believed that there was a chance that angled endoscopes may be sufficient not only to see, but also to guide the microsurgical dissection of the foraminal tumor. If this can be accomplished, it would obviate the need for the drilling of the foramen. As such, we designed for this patient an endoscopic-assisted, retrosigmoid keyhole approach for removing his schwannoma.

■ Description of the Technique

After establishment of general anesthesia and placement of monitoring electrodes for somatosensory evoked potential and facial nerve recording, the patient was positioned lateral, with the neck flexed but neutral in rotation. A 2.5-inch incision in

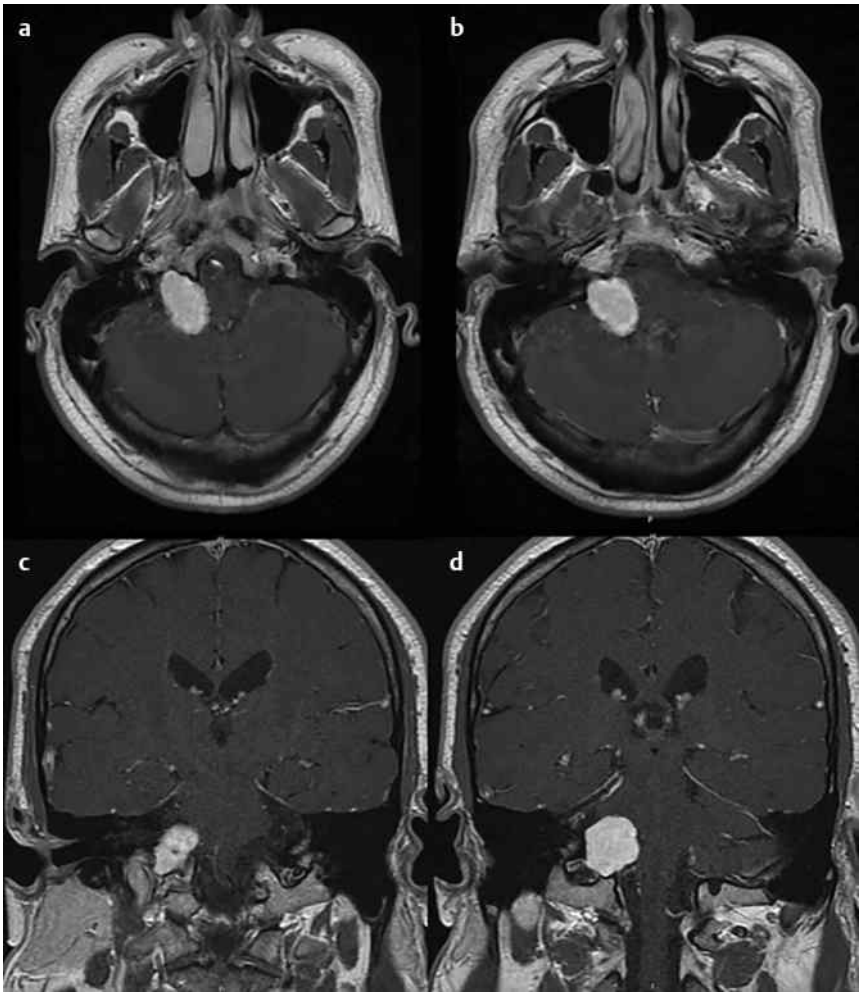


Fig. 20.7 Preoperative MRI. (a, b) Axial and **(c, d)** coronal images after gadolinium administration showing a jugular foramen tumor consistent with a schwannoma. The tumor is mainly intradural, but there is a significant component in the bony foramen, most clearly demonstrated in **(a)** and **(c)**.

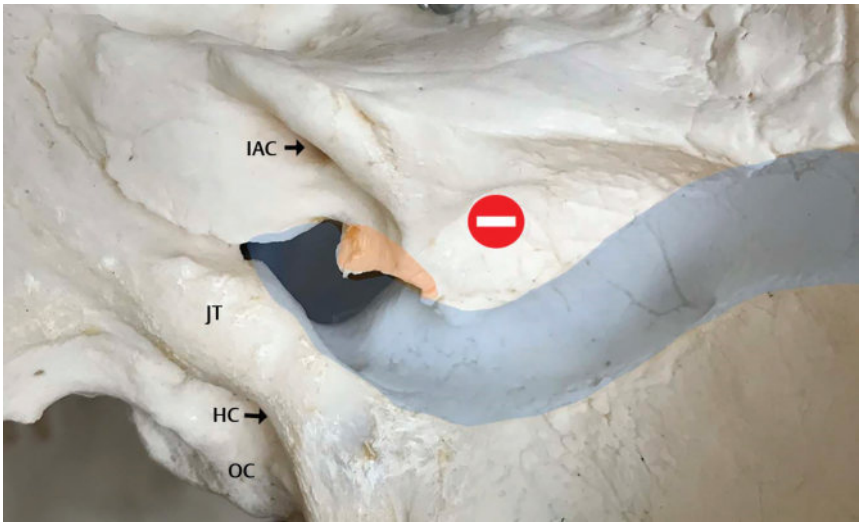


Fig. 20.8 Dry skull image of the right jugular foramen. The area in orange is safe for removal to improve access to the tumor in the bony foramen. This corresponds to the area removed during the "suprajugular" or "infralabyrinthine" approaches. The "no entry" sign overlies the area of the endolymphatic sac, and care must be taken that drilling stops before reaching this area. HC, hypoglossal canal; IAC, internal acoustic canal; JT, jugular tubercle; OC, occipital condyle.

the shape of a "lazy S" was made two fingerbreadths behind the right ear, starting at the level of the transverse sinus. A burr hole was made just below the asterion, and a 1.5-inch diameter keyhole craniotomy was created posteroinferior to this burr hole. Using a rounded burr, bone was removed at the anterior margin of the craniotomy up to the posterior edge of the sigmoid sinus. The dura was cut and reflected anteriorly. The drill was fitted with a 2-mm diamond burr, ready to be used for the drilling at the jugular foramen.

Intradurally, CSF was evacuated from the cisterna magna to relax the cerebellum. The cerebellum was then mobilized posteriorly to expose the cerebellopontine angle, where the bulk of the tumor was visualized under the microscope (**Fig. 20.9a**). As the tumor was relatively avascular, the capsule of it was opened to

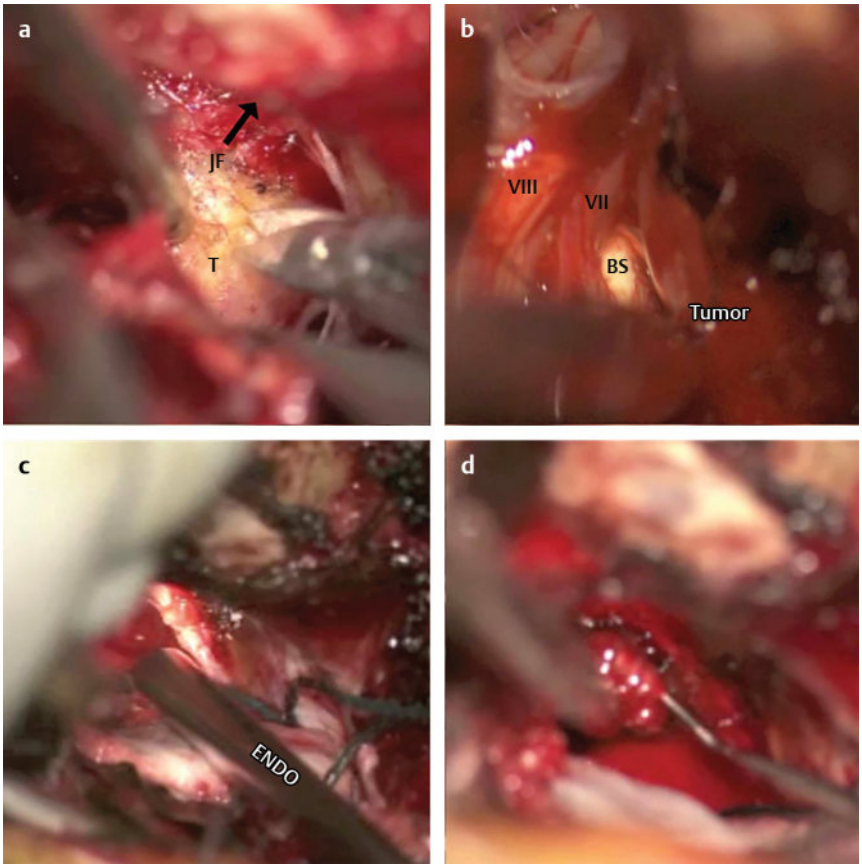


Fig. 20.9 Intraoperative images. (a) The tumor was exposed in the cerebellopontine angle (*black arrow*). JF, jugular foramen; T, tumor. **(b)** After removing the bulk near the superior pole, the capsule is dissected away from the cranial nerves nearby. BS, brainstem; VII, facial nerve; VIII, vestibulocochlear nerve. **(c)** After removing all the intradural portion of the tumor, the endoscope was utilized to dissect the foraminal component, to loosen adhesions and bring pieces closer to the foraminal opening. **(d)** Curved dissectors were used to remove the final pieces of the tumor inside the foramen.

allow debulking with the tumor aspirator. The removal of some of the bulk allowed mobilization of the inferior capsule away from the brainstem, and once this plane was protected with a cottonoid, the debulking resumed near the upper pole of the tumor. The dissection of the upper capsule was then performed with the specific goal of freeing the facial and vestibulocochlear nerves from adhesions to the capsule (**Fig. 20.9b**). The neural integrity monitor was used liberally to assist in this maneuver. Once freed, these cranial nerves were padded away with cottonoids.

The tumor removal continued at this point until the intradural component was completely removed. The 30-degree endoscope was then inserted under microscopic guidance and from here on, the operation alternated between endo- and microscopic visualization (**Fig. 20.9c**). With the endoscope in the nondominant hand and curved dissection and resection tools in the dominant one, some intraforaminal tumor pieces were mobilized and removed. To avoid excessive traction forces, other more adherent pieces were loosened under endoscopic visualization and mobilized close to the opening, but then removed with curved forceps using two-hand technique back under the microscope (**Fig. 20.9d**). By alternating between endo- and microscopic visualization, the foraminal component of the tumor was removed completely, obviating the need for further widening of the foramen with drilling.

A short video demonstrating the technique accompanies this chapter (**Video 20.1**).

■ Aftercare

After surgery, the patient experiences significant swallowing problems. This was not unexpected given his preoperative symptoms and fortunately, his swallowing function returned to normal with time. The postoperative MRI scan showed a complete removal of the tumor, and at the 30-month follow-up, the patient remained tumor- and symptom free (**Fig. 20.10**).

■ Commentary

Regarding trends in the jugular foramen literature, another thread that ran through most of the publications was the importance in tailoring the operation to fit the particular configuration of the individual patient's tumor. Depending on the classification one uses, there are only either three or four types of jugular foramen schwannoma, but as all the experts stressed, there are far more subtle variations in these tumors which can affect surgical choices. Tumors that differ subtly in anatomy may require dramatically different approaches, and therefore, it would be a misleading simplification to suggest a correlation between a tumor type and approach for resection.

For schwannomas that are more or less "dumbbell-shaped" or have a large extracranial component, larger "skull base" approaches with full opening of the foramen and elimination of the jugular system would be necessary to remove the whole tumor. What this chapter has shown is that intradural jugular foramen tumor with foraminal extension can be resected fully without giant transjugular operations. For the operation in this section, we were able to completely remove the tumor using a keyhole approach, assisted by angled endoscopes and curved dissection tools. It must be stressed that we were fully prepared for intradural, suprajugular drilling, as there was no way to predict preoperatively how the tumor would mobilize out of the

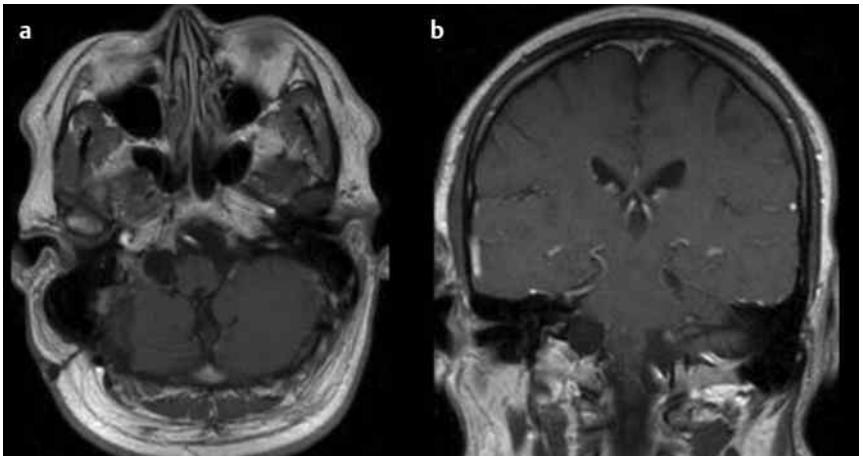


Fig. 20.10 Postoperative MRI. (a, b) A follow-up study done at 30 months after the endoscopic-assisted, retrosigmoid keyhole approach demonstrated complete tumor removal without recurrence.

foramen. Had the tumor been more adherent or the visualization with the endoscope been suboptimal, we would have been ready to open the foramen as one would the internal acoustic meatus during vestibular schwannoma surgery. Indeed, even though meticulous preoperative planning can tailor the operation to a certain degree, intraoperative assessment and adjustments are frequently necessary to fine-tune the necessary steps of the operation.

Our example once again highlights the importance of endoscopy in modern skull base surgery. We knew that a large transjugular operation was not necessary to remove the patient's schwannoma, but with the help of the endoscope, we were able to avoid any drilling of the foramen altogether.

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21 Jugular Foramen (Intra- and Extracranial)

Alexander Tai, R. Tushar Jha, Walter C. Jean, and Amjad Anaizi

Keywords: paraganglioma, glomus jugulare, infralabyrinthine approach, facial nerve

■ Case Presentation

A 42-year-old man presented to the hospital with the chief complaint of worsening headache, nausea, and vomiting. He was a patient of the neurosurgical clinic, with a long history of a glomus jugulare tumor. His first surgery was 16 years prior to the current presentation at another institution, and it involved a posterior craniocervical, transmastoid, transpetrosal approach with closure of his ear canal. Because of significant blood loss at that time, the operation had to be truncated, and the postoperative residual tumor was treated with 45 Gy in 25 fractions of radiotherapy. He had subsequently undergone two additional partial resections for persistent growth of the tumor and brainstem compression, and his other surgeries included placement and removal of a ventriculoperitoneal shunt for infection. His last operation was 5 years prior to the current presentation.

A recent magnetic resonance imaging (MRI) showed that the tumor was once again growing, effacing the fourth ventricle and compressing the brainstem. He had presented to clinic with headaches, gait instability, and difficulty swallowing. Treatment options had been presented to the patient and his family, and it was during the deliberation of the plans that his headaches worsened to a point where he sought emergent treatment.

On examination, he was awake and fully oriented, with a stable House–Brackmann III left-sided facial palsy, which he had had for 16 years. He had no hearing on the left. His speech was hoarse and the tongue deviated to the left. He was dysmetric in his left upper extremity. Atrophy of his trapezius and sternocleidomastoid (SCM) muscles were noted on visual inspection. The computed tomography (CT) scan ordered by the emergency department showed stable size and mass effect of his lesion and a "moth-eaten" pattern of destruction of the adjacent temporal bone. It also revealed a new third ventricular hemorrhage without associated hydrocephalus (**Fig. 21.1**). He was transferred to the University Hospital neurosurgical intensive care unit (ICU) for further management. On arrival, a new MRI of the brain with and without contrast was obtained (**Fig. 21.2**).

Questions

1. What is the differential diagnosis of tumors at the jugular foramen?
2. What further labs/imaging/tests should be included in work-up of a glomus jugulare tumor?
3. Are there potential genetically based markers/syndromes associated with glomus jugulare tumors?

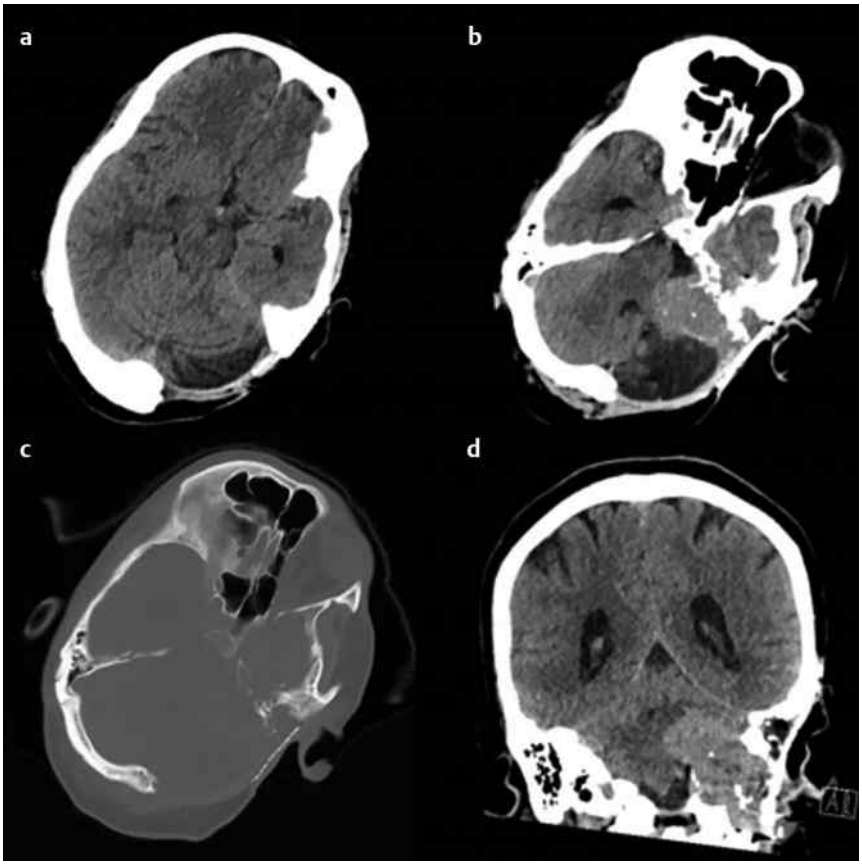

Fig. 21.1 Preoperative CT. Images of the patient's CT on presentation to the emergency department with axial cuts demonstrating **(a)** hemorrhage in the third ventricle and **(b)** a jugular foramen tumor extending superiorly and medially compressing the brainstem. **(c)** Bone windows demonstrated a previous suboccipital craniectomy and mastoidectomy likely from retrosigmoid and presigmoid approaches to the tumor. **(d)** Coronal slices demonstrated the extent of tumor infiltration into the petrous bone.

Diagnosis and Assessment

Laboratory examination for the patient included urinary tests for vanillylmandelic acid and metanephrines. These tests were negative, indicating that the tumor was nonsecretory. The importance of knowing the secretory status of paragangliomas cannot be overemphasized, as preoperative sympathetic blockade is frequently needed to prevent intraoperative hypertensive crisis for secretory tumors. Audiological testing is of course a routine part of the examination for patients with jugular foramen tumors, but because this patient had a closed ear canal, it was omitted. Bedside endoscopy was performed to confirm that his left vocal cord was paralyzed. His family history was unremarkable for familial paraganglioma syndromes, and previous genetic work-up was negative for multiple endocrine neoplasia type 2, neurofibromatosis type 1, or von Hippel–Lindau syndrome.

The patient's MRI showed a large tumor with brainstem compression and fourth ventricular effacement. The lesion enhanced homogeneously, occupied the jugular foramen and extended into the cerebellopontine angle (CPA), anterior to the brainstem, up the clivus into the cavernous sinus. In the extracranial compartment, it also extended along the floor of the middle fossa into the infratemporal fossa and neck (**Video 21.1**). The internal carotid artery (ICA) was encased by tumor from the cervical to cavernous segment. The vertebral artery coursed inferior and anterior to the lesion. On T2-weighted imaging, the lesion demonstrated a "salt-and-pepper" appearance representing small foci of previous hemorrhage (**Fig. 21.2**). This appearance is typical of glomus tumors. Imaging artifact was noted in the overlying soft tissue indicative of prior embolization.

Cerebral angiography demonstrated no flow in the left sigmoid sinus and internal jugular vein (IJV), as expected from the patient's history of a glomus tumor as well as his previous surgeries. On the other hand, there was extensive tumor "blush" with feeders directly from the left vertebral artery, left posterior inferior cerebellar, left anterior inferior cerebellar artery, left external carotid artery (ECA) (including the left maxillary, middle meningeal, and lingual arteries), left ICA, and even the left ophthalmic artery (**Fig. 21.3**). There was evidence

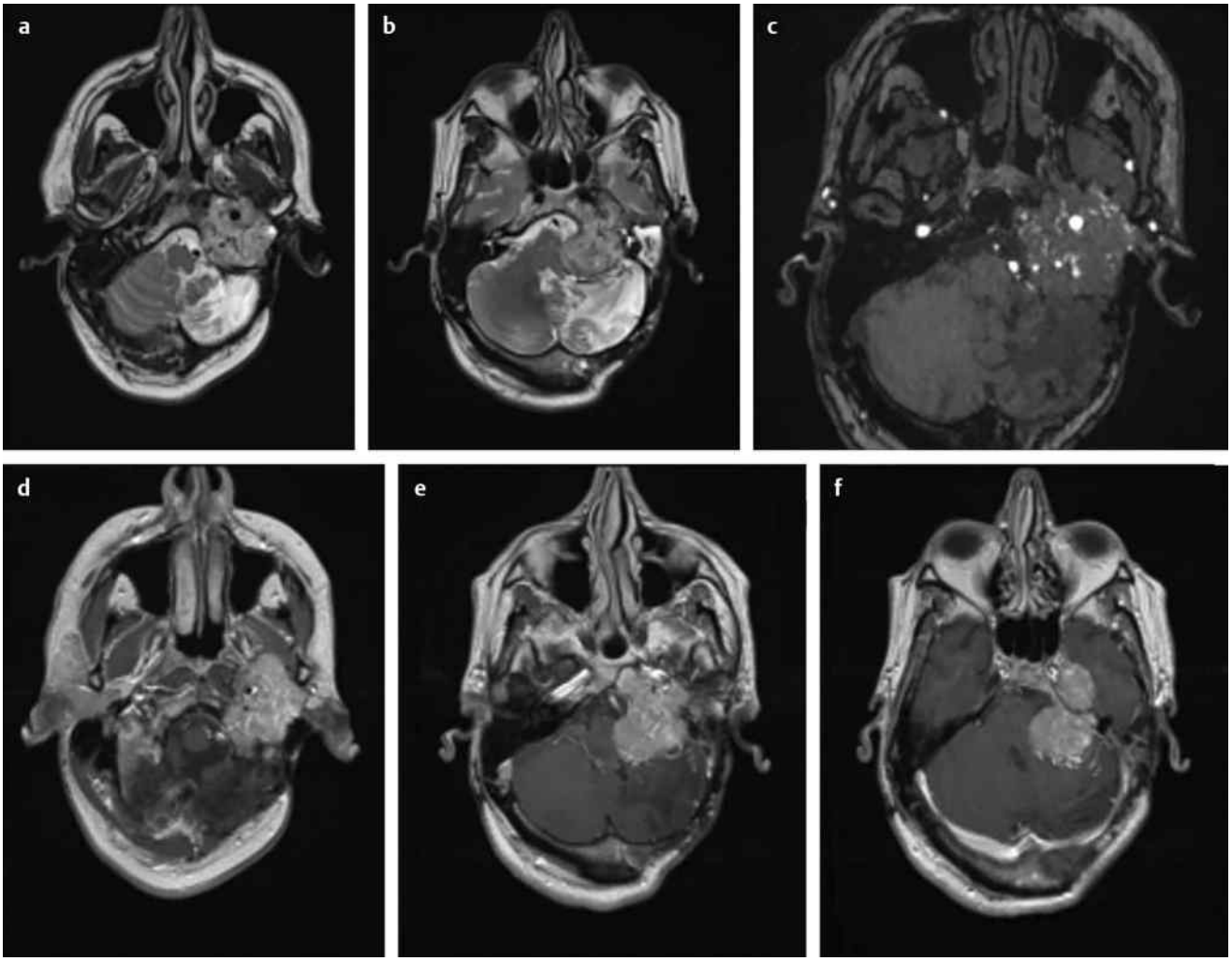

Fig. 21.2 Preoperative MRI. These images of the patient's jugular foramen tumor showed **(a)** the "salt-and-pepper" appearance of the tumor representing significant vascularity and **(b)** the degree of brainstem compression as it extended medially. **(c)** Time-of-flight MR angiography demonstrated that the cervical ICA is completely encased in the tumor. **(d)** Axial MR T1 postcontrast cuts demonstrated the tumor's extensive infiltration into the infratemporal fossa, **(e)** posterior fossa and **(f)** middle fossa.

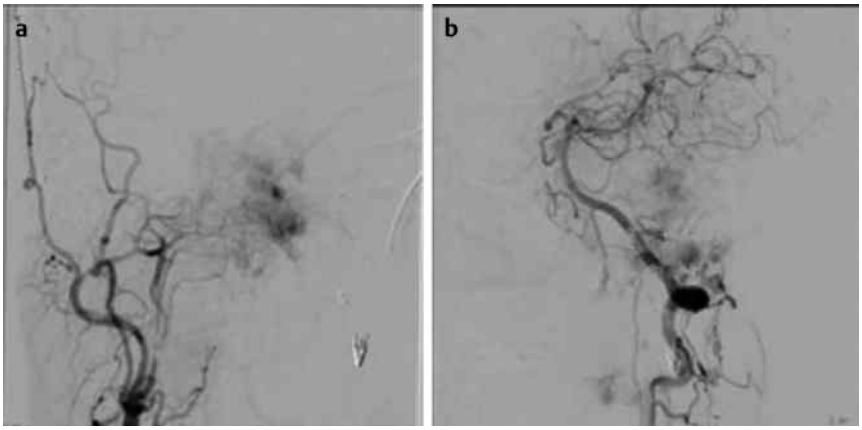

Fig. 21.3 Preoperative angiogram. **(a)** Anteroposterior external carotid and **(b)** lateral vertebral artery injections demonstrated the tumor's multiple arterial feeders.

of previous embolization performed in the branches of the left ECA. Indeed, coils were visible in the left occipital and middle meningeal arteries. Despite this, the tumor seemed to have recruited feeders from the various large vessels which were mentioned earlier, and these feeders were all tiny in size.

The differential diagnosis for a jugular foramen tumor includes paragangliomas, schwannomas, meningiomas, chordomas, chondrosarcomas, and metastatic tumors. The typical "salt-and-pepper" appearance on CT and MRI, along with the tumor invasion into the neck, made the diagnosis of a glomus jugulare quite straightforward in this patient, even if one had inspected his images without information from his previous surgeries. Newly diagnosed glomus jugulare tumors are treated with stereotactic radiosurgery with increasing frequency. The main reasons for this are twofold: radiosurgery is noninvasive and quite effective for small paragangliomas, and surgery for these lesions, even small ones, involves complex approaches with significant morbidities.

However, the large size of this patient's tumor, along with his history of previous radiation treatment, ruled out radiotherapy as a stand-alone treatment for his current problem. His progressively worsening symptoms were caused by increasing brainstem compression, and thus surgery was undoubtedly necessary. However, aggressive removal of the tumor that goes beyond the goal of brainstem decompression would involve a massive operation, as the lesion not only invaded all the way anteriorly to the pterygoid fossa, but also encased the ICA from the cervical to cavernous segment (**Fig. 21.4**). Before we determine if the morbidity of this is justifiable, we must consider the various treatment options.

■ Anatomical and Therapeutic Considerations

The jugular foramen is formed anteriorly by the petrous portion of the temporal bone and posteriorly by the occipital bone. It is located rostral and lateral to the hypoglossal canal and caudal and medial to the internal acoustic meatus. In most patients, the right jugular foramen is larger than the left. It is canonically divided into two compartments, the anteromedial pars nervosa and posterolateral pars venosa. Often these two compartments are separated by a thin bony septum, the jugular spine. Pars venosa contains the jugular bulb, the posterior meningeal branch from the ascending pharyngeal artery,

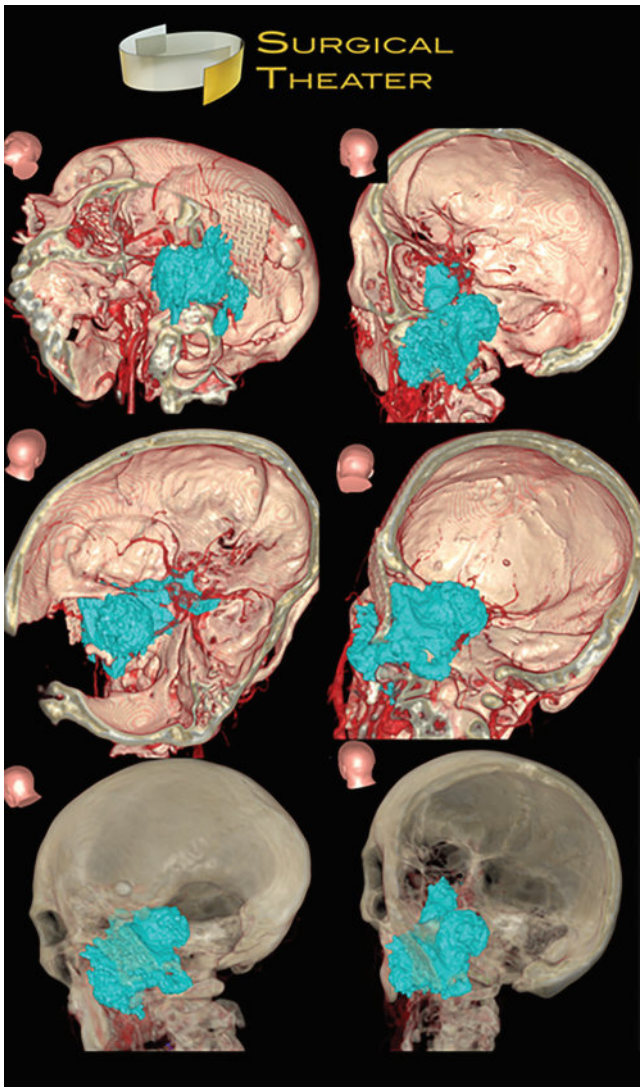

Fig. 21.4 Three-dimensional rendering of the skull base. The head position for each image is shown in the upper left corner. This massive recurrent glomus jugulare tumor occupied the entire left posterolateral skull base, compressing the brainstem anteriorly from the cerebellopontine angle to the prepontine cistern. It encased the internal carotid from its cervical to cavernous segment and filled the cavernous sinus. Extracranially, it extended along the middle fossa floor into the infratemporal and pterygoid fossa. The vertebral artery coursed inferior and anterior to the lesion.

and cranial nerves X and XI. Pars nervosa contains the inferior petrosal sinus, several meningeal branches from the occipital artery and cranial nerve IX. As mentioned in the previous chapter, the most recent data indicate that there may in fact be three compartments, one with cranial nerve IX, another with cranial nerves X and XI coursing with a common dural sheath, and a third, "interperiosteodural" compartment with blood from the inferior petrosal sinus and sigmoid sinus. Lastly, an important anatomic consideration when treating glomus jugulare tumors is that they arise from the chief cells of the paraganglia located within the wall of the jugular bulb and can be associated with either Arnold's nerve, the auricular branch of cranial nerve X located in pars venosa, or Jacobson's nerve, the tympanic branch of cranial nerve IX located in pars nervosa.

Options for Approach

When planning to treat glomus tumors, it is critical to keep in mind the goals of therapy. Most glomus tumors have a relatively slow growth rate and over time can infiltrate deeply into the skull base. With this consideration in mind, a gross total resection may not always be the best therapy for all patients. In fact, many small lesions may be observed or managed with stereotactic radiosurgery. When dealing with large tumors, skull base infiltration can complicate resection, as multiple surgical corridors are needed to address intracranial and extracanial portions of the tumors.

Contemporary approaches to glomus jugulare tumors owe most of their foundation to the groundbreaking work done by Ugo Fisch in the 1970s. The Fisch type A infratemporal fossa approach remains the basis on which all contemporary approaches to the jugular foramen are built. The basic elements of the approach include a transmastoid/transpetrosal exposure of the sigmoid sinus and jugular bulb, neck dissection to expose and control the cervical carotid, IJV and lower cranial nerves, ligation of the sigmoid sinus and IJV, and finally opening the venous structures to remove the tumor and packing the inferior petrosal sinus openings (**Fig. 21.5**).

This approach was designed to target tumors in the sigmoid sinus, jugular bulb, and medial to the facial nerve, while its "sibling approaches," the Fisch types B and C, were designed for tumors that are progressively more anterior and deep, surrounding the petrous carotid in the posterior infratemporal fossa and invading further into the pterygopalatine fossa (**Fig. 21.6**, **Fig. 21.7**). While all three approaches involve the surgical closure of the external auditory canal, and therefore, hearing loss, the anterior mobilization of the facial nerve out of the fallopian canal may not be universally necessary (shown here only with type A, **Fig. 21.5**).

As surgeons gained experience with approaches to the jugular bulb area, they began to put a premium on the preservation of cranial nerve function, and the thought of sacrificing intact functions to gain exposure became progressively more onerous. The development of the "fallopian bridge" was an example of this "concept evolution." With this technique, the facial nerve is no longer taken out of its canal and moved anteriorly. Instead, the bone around the fallopian canal is drilled away until a thin rim of it is left around the facial nerve, and neither the nerve nor the cylinder of bone around it is touched during the rest of the operation, which takes place in front and behind this "bridge."

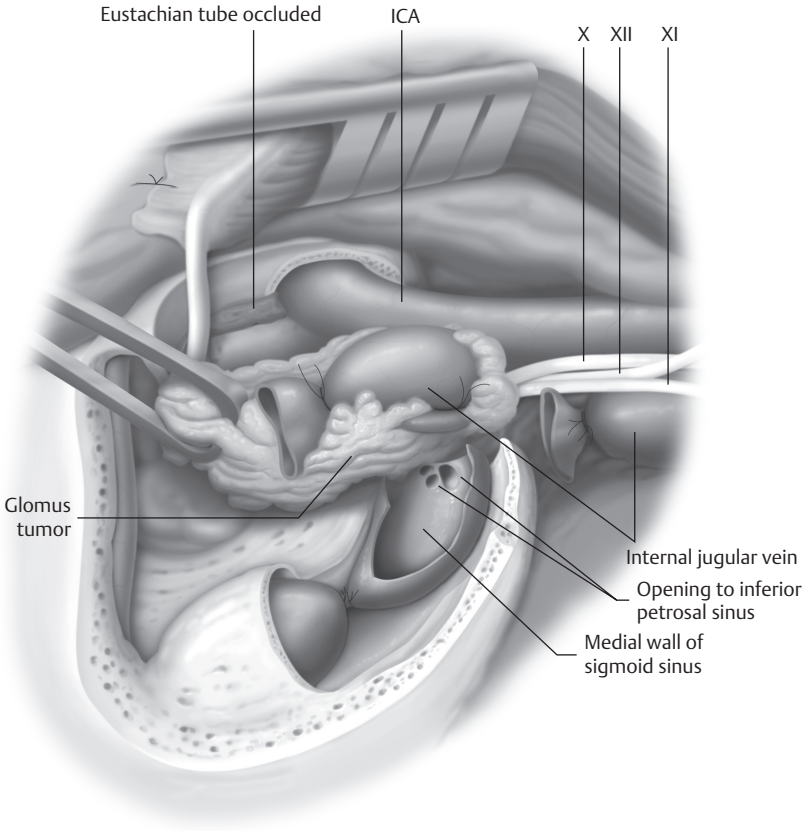

Fig. 21.5 Fisch type A (right side). Illustration depicting the exposure of the Fisch infratemporal fossa approach, type A. Note that the ear canal is surgically closed and the facial nerve is transposed anteriorly. ICA, internal carotid artery; X, vagus nerve; XI, accessory nerve; XII, hypoglossal nerve. (Reproduced from Friedman R, Slattery III W, Brackmann D et al, eds. Lateral Skull Base Surgery. The House Clinic Atlas. 1st ed. Thieme; 2012.)

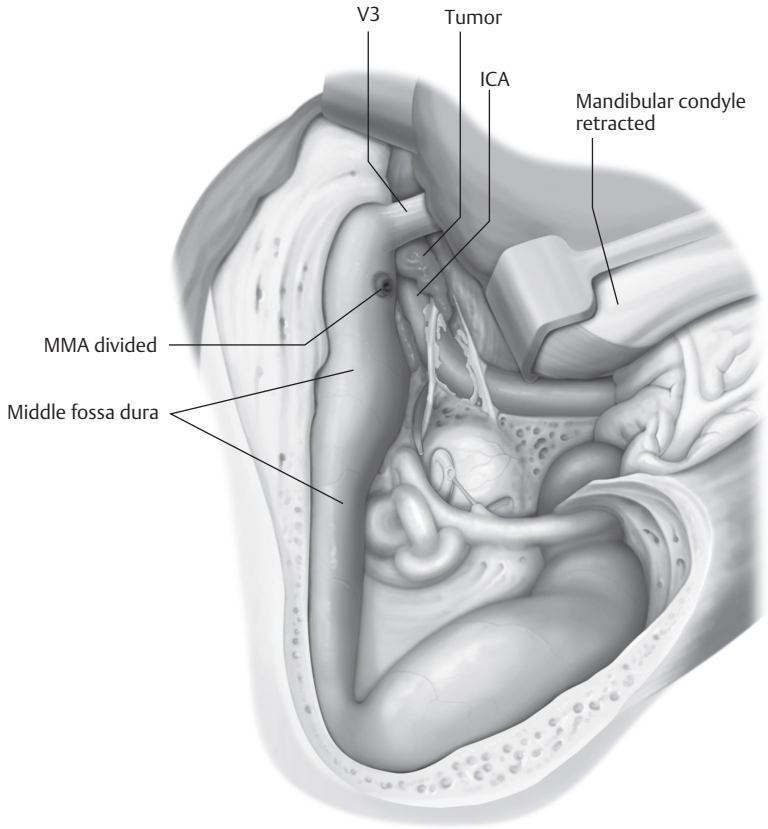

Fig. 21.6 Fisch type B, right side. Illustration depicting the exposure of the Fisch infratemporal fossa approach, type B. The horizontal intrapetrous carotid artery is exposed to gain safe access to the petrous apex and clivus. This approach requires removal of the glenoid fossa, division of the middle meningeal artery (MMA), and division of V3. ICA, internal carotid artery. (Reproduced from Friedman R, Slattery III W, Brackmann D et al, eds. Lateral Skull Base Surgery. The House Clinic Atlas. 1st ed. Thieme; 2012.)

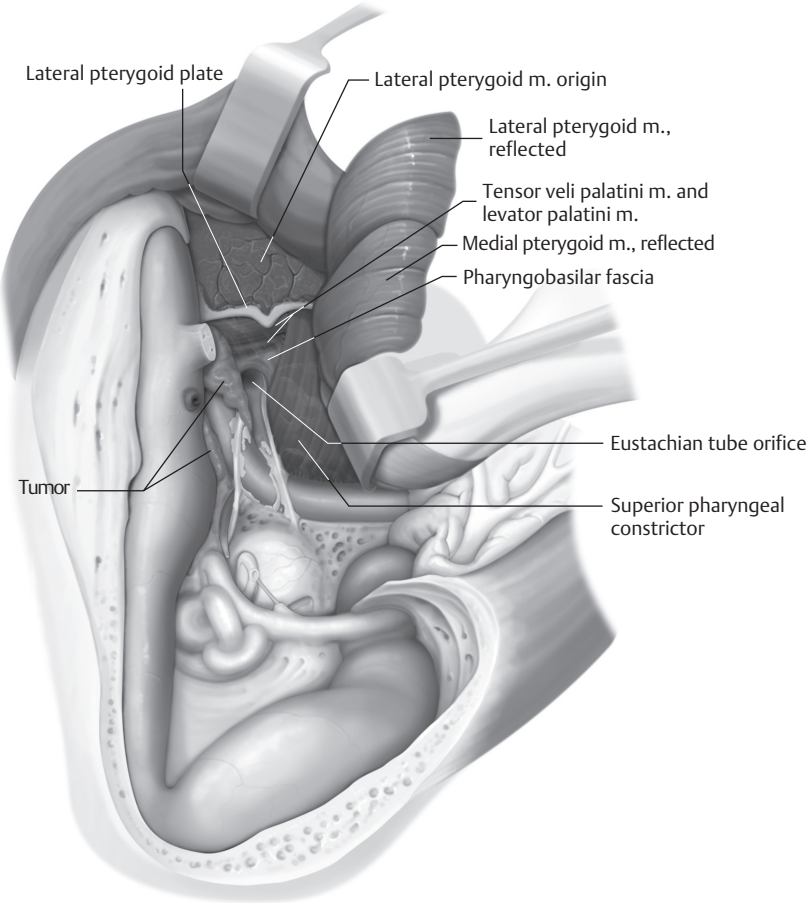

Fig. 21.7 Fisch type C, right side. Illustration depicting the exposure of the Fisch infratemporal fossa approach, type C. This allows access to the pterygopalatine fossa, the nasopharynx, and the cavernous sinus. (Reproduced from Friedman R, Slattery III W, Brackmann D et al, eds. Lateral Skull Base Surgery. The House Clinic Atlas. 1st ed. Thieme; 2012.)

From here, the concept evolution followed the trend, which was detailed in this book's Introduction, of considering approach elements, such as closure of the ear canal or facial nerve mobilization, as "building blocks," that can be added or subtracted from a basic platform, in order to tailor the approach to a specific patient and tumor. The modern-day operation for glomus jugulare tumors has a basic platform with the essential approach and resection steps as follows:

1. Large C-shaped incision behind the ear and a neck incision following the contour of the SCM muscle (**Fig. 21.8**).

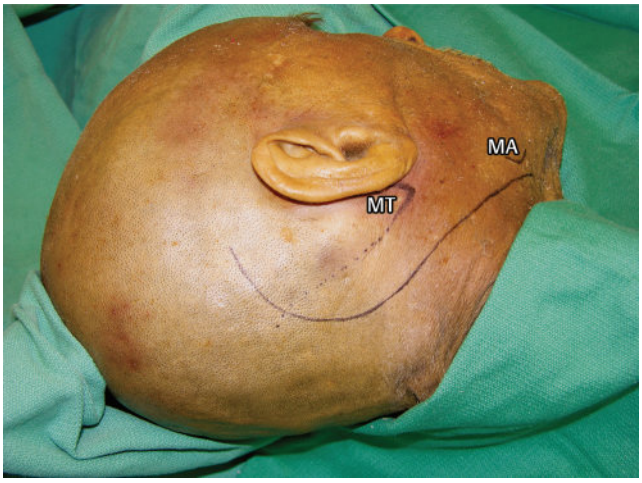

Fig. 21.8 Cadaveric dissection demonstrating the approach for a glomus jugular tumor. With the patient in the lateral position, a "C" shaped incision is made that begins above and behind the ear and extends below the angle of the mandible. The skin incision extends posterior to the asterion to expose the mastoid portion of the occipital bone and inferiorly below the angle of the mandible to expose the internal jugular vein. MA, mandibular angle; MT, mastoid tip. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

2. Mobilization of the SCM in a direction of the surgeon's preference; dissection of the IJV, ICA, and cranial nerves IX, X, XI, XII in the neck (**Fig. 21.9**).
3. Detaching the posterior belly of the digastric muscle off the mastoid and retracting it anteriorly.
4. Mastoidectomy; skeletonizing the sigmoid sinus and jugular bulb (**Fig. 21.10**).
5. Ligation of the sigmoid sinus, partial occlusion of the IJV, opening of the sigmoid sinus for tumor removal (**Fig. 21.11**).
6. Ligation of IJV, completion of extradural tumor removal, with preservation of the anterior wall of the jugular bulb and IJV for cranial nerve protection.
7. Plugging of the inferior petrosal sinus openings, and elimination of the posterior condylar vein.
8. Opening the dura for intradural tumor removal (**Fig. 21.12**).

Essentially a "transmastoid, infralabyrinthine approach," an operation with these steps is suitable for tumors in the jugular bulb, upper neck, with limited intradural extension, and blood supply exclusively from the external carotid branches. For tumors with more extensive intradural extension, especially ventrally toward the clivus, the approach elements of occipital condylectomy, drilling the jugular tubercle and unroofing the hypoglossal canal can be added to increase the exposure.

For bigger tumors, more elements would be added to the basic platform. A tumor with ICA blood supply from its cervical segment would necessitate exposure anterior to the facial nerve. Here, the fallopian bridge technique would prove handy, and the resection would employ the two corridors in front and behind the bridge. Still bigger tumors that involve or encase not only the cervical but petrous and lacerum segments of the ICA, reaching the cavernous sinus, would require the additional closure of the external acoustic canal for exposure. Patients with even bigger tumors are likely to have little to no preoperative facial nerve function. For these patients, the anterior transposition of the facial nerve is justifiable to gain wide surgical exposure.

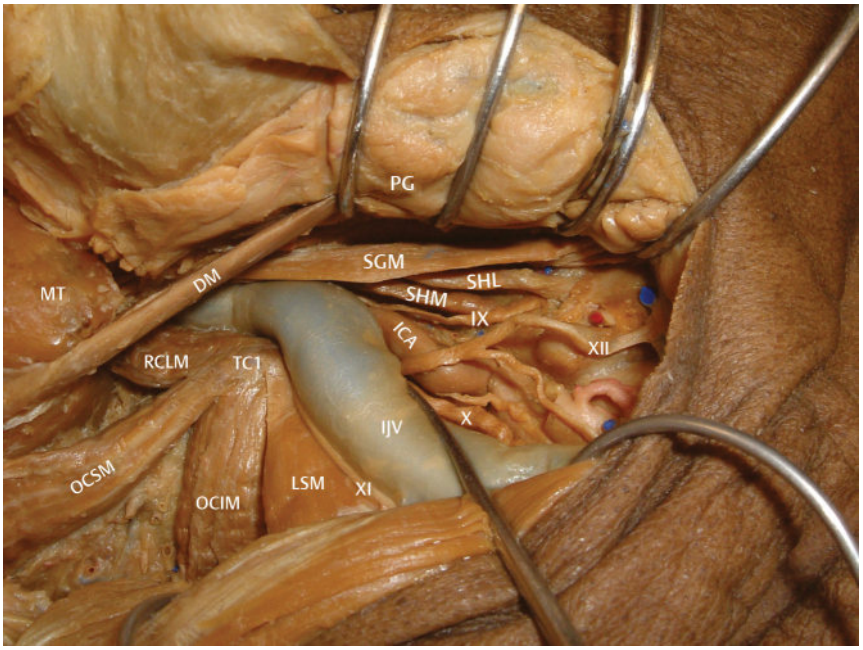

Fig. 21.9 Cadaveric dissection: Overview of the cranial nerves. The hypoglossal nerve is seen passing under the greater horn of the hyoid bone. The vagus nerve is seen between the internal carotid artery and internal jugular vein. The glossopharyngeal nerve is seen under the stylohyoid muscle and the spinal accessory nerve is seen under the lateral process of C1. DM, digastric muscle; ICA, internal carotid artery; IJV, internal jugular vein; IX, glossopharyngeal nerve; LSM, levator scapulae muscle; MT, mastoid tip; OCIM, obliquus capitis inferior muscle; OCSM, obliquus capitis superior muscle; PG, parotid gland; RCLM, rectus capitis lateralis muscle; SGM, styloglossus muscle; SHL, stylohyoid ligament; SHM, stylohyoid muscle; TC1, transverse process of C1; X, vagus nerve; XI, accessory nerve; XII, hypoglossal nerve. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st Ed. Thieme; 2009.)

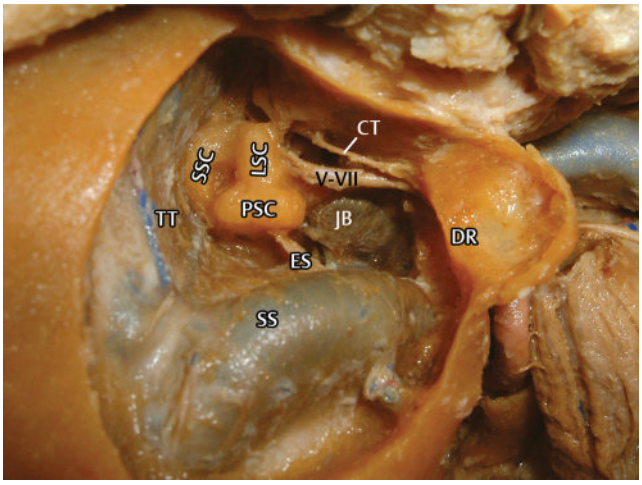

Fig. 21.10 Cadaveric dissection: Mastoidectomy, exposing the jugular bulb. The facial nerve is dissected from the surrounding bone as is the chorda tympani. The facial nerve is first defined anterior to the lateral semicircular canal. The nerve is then followed inferiorly to the styloid foramen. The jugular bulb is found inferior and deep to the semicircular canals. Its size is variable. Facial nerve decompression should be performed slowly and carefully. This procedure should not be hurried. The sharp bony notch between the inferoanterior border of the sigmoid sinus and the posterior border of the jugular bulb may be very adherent to the vein. It is often best to thin this bone and leave the pliable thin bone in place. CT, chorda tympani; DR, dural ring; ES, endolymphatic sac; JB, jugular bulb; LSC, lateral semicircular canal; PSC, posterior semicircular canal; SS, sphenoid sinus; SSC, superior semicircular canal; TT, temporal tegmen; V–VII, vertical segment of the facial nerve. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

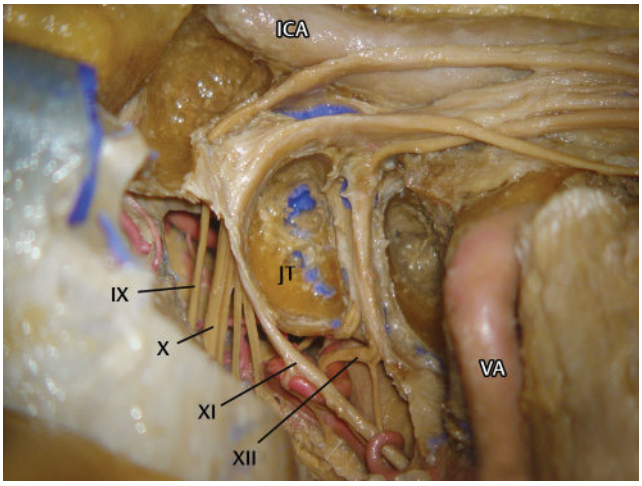

Fig. 21.12 Cadaveric dissection: Intradural exposure of the jugular foramen. The jugular tubercle has been partially removed to increase exposure. Note the right angle turn the hypoglossal nerve makes as it exits the hypoglossal canal. ICA, internal carotid artery; IX, glossopharyngeal nerve; JT, jugular tubercle; VA, vertebral artery; X, vagus nerve; XI, accessory nerve; XII, hypoglossal nerve. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

Approach of Choice

Our patient's previous surgeries have included a transpetrosal, transmastoid approach with closure of the external auditory

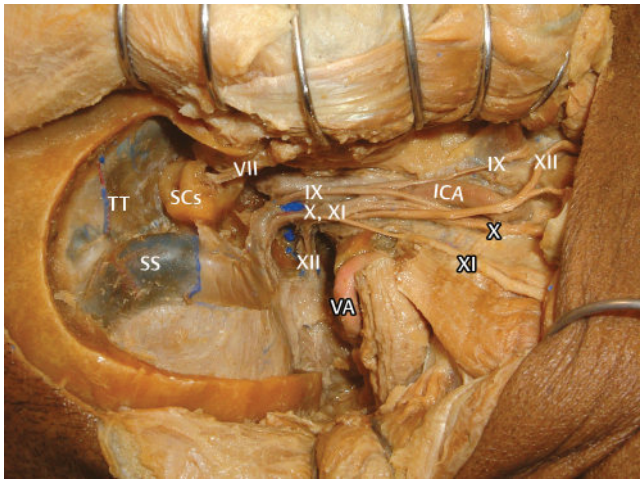

Fig. 21.11 Cadaveric dissection: Ligation of the sigmoid sinus and opening of sinus for tumor removal. Most glomus jugulare tumors are extradural and reside in venous structures. This allows the surgeon to remove the lateral portion of the sigmoid sinus, leaving the medial wall, which abuts the cranial nerves, intact. Here the internal jugular vein has been removed to demonstrate the anatomy behind it. ICA, internal carotid artery; IX, glossopharyngeal nerve; SCs, semicircular canals; SS, sphenoid sinus; TT, temporal tegmen; VA, vertebral artery; VII, facial nerve; X, vagus nerve; XI, accessory nerve; XII, hypoglossal nerve. (Reproduced from Wanibuchi M, Friedman AH, Fukushima T, eds. Photo Atlas of Skull Base Dissection: Techniques and Operative Approaches. 1st ed. Thieme; 2009.)

canal. In addition, because of the intradural component of the tumor, a far-lateral craniotomy with partial condylectomy was performed (**Fig. 21.2**). The goal of the original surgery was to achieve a gross total resection; however, significant intraoperative blood loss was encountered, forcing the surgery to be aborted. Further attempts at resection, preceded by endovascular embolization, were again limited due to blood loss. Despite these operations, his facial nerve function was graded at HB3, and his extraocular motility was intact even though the cavernous sinus was filled with tumor.

For such a complex patient, the deliberation of options and establishment of the surgical goal is best done before a multidisciplinary panel. With subspecialists from the fields of endovascular therapy, radiosurgery, otolaryngology, and neurosurgery present, the group acknowledged that total removal of the tumor would be highly risky given that several previous operations had been aborted because of massive blood loss. As large feeding vessels had been blocked by coils previously deployed, catheter access to the tumor for reliable and effective embolization to reduce the blood supply was no longer feasible. The nutrient vessels to the tumor, though numerous, were all too small for endovascular access. Although alternatives such as direct puncture of the tumor may work, the results are far from reliable.

Since the patient's oculomotility remained intact, it was reasonable to consider leaving tumor untouched in the cavernous sinus. The encased ICA can potentially be bypassed with a cervical to supraclinoid graft, but surgical exenteration of the cavernous sinus would leave the left eye unusable. If the cavernous sinus portion of the tumor can rationally be left for further radiation treatment, might it not be reasonable also to leave other portions for the same? Since the previous radiotherapy was more than a decade ago, the multidisciplinary panel agreed that new treatment with radiosurgery was

feasible if the target size was significantly reduced, and with that, a rational surgical goal seemed to emerge.

The patient was primarily experiencing signs and symptoms of progressive brainstem compression. While the anterolateral portions of the lesion were significant, they were not an immediate threat to the patient. If a posterolateral approach through the patient's previous far-lateral and transpetrosal exposure was successful in decompressing the brainstem and reducing the tumor volume, then postoperative radiosurgery might control the remaining portions.

Questions

1. The tumor extended anterior to the brainstem. What maneuvers may increase exposure and provide increased access to this region?
2. Assuming that the posterior fossa tumor could be removed surgically, what dose of radiosurgery (SRS) would be reasonably used to treat the rest of the tumor?

Description of the Technique

For the first stage of treatment, the patient was taken to the angiography suite for direct-puncture embolization of the tumor (**Fig. 21.13**). The patient was positioned supine with the head turned to the right. Neuronavigation was registered with excellent accuracy using an array attached to a headband. A 20-gauge spinal needle was then registered with the neuronavigation system. A safe retrosigmoid trajectory to the tumor was planned. The spinal needle was slowly passed through the titanium mesh and inserted into the tumor using neuronavigation and fluoroscopy for simultaneous guidance. Contrast was then injected through the spinal needle confirming a tumor blush. A CT was also done to confirm satisfactory location of the needle within the tumor. Onyx was then injected through the spinal needle under angiographic visualization achieving successful partial embolization of the tumor. A total of 12.8 mL of Onyx was injected into the tumor over 28 minutes. This achieved an approximate 75% reduction in the vascular supply to the tumor. The plan was to obtain an updated MRI that evening and proceed with open surgical resection the following morning.

In the operating room, the patient was positioned in the lateral position with head secured in Mayfield pins. Intraoperative neuronavigation was registered with very good accuracy. A portion of the previous retroauricular incision was incised. The titanium mesh cranioplasty was removed revealing the transpetrosal/far-lateral exposure. The dura was opened in a T-shaped fashion. Cerebrospinal fluid (CSF) drainage from the cisterna magna achieved excellent cerebellar relaxation and the tumor capsule was identified. The mass was then internally debulked with the ultrasonic aspirator, removing tumor as well as significant amounts of Onyx. With significant removal of tumor bulk, the medial edge of the capsule was then gently mobilized off the cerebellum and brainstem. The vertebral artery, which coursed medial to the lesion, was visualized only after a majority of the posterior fossa tumor was resected. Once the inferior pole of the intradural portion of the tumor was visualized, resection continued superiorly (**Fig. 21.14**).

The tumor was visualized extending through the tentorium. The middle fossa component of the tumor was then accessed through the transtentorial corridor created by the tumor. Prior to extending the tentorial opening, it became clear that the middle fossa component of the tumor was not well penetrated with Onyx, as rapid and voluminous blood loss limited the superior extent of the resection. No further attempts were made to access the supratentorial extent of the tumor. The portions of the lesion extending into the infratemporal fossa and neck were left untouched. At the end of the operation, the brainstem was clearly visualized and well decompressed (**Fig. 21.15**). The dura was closed in watertight fashion. A titanium mesh was used to reconstruct the bony defect and the fascia and skin closed in anatomic layers. Intraoperative blood loss was 1500 cc.

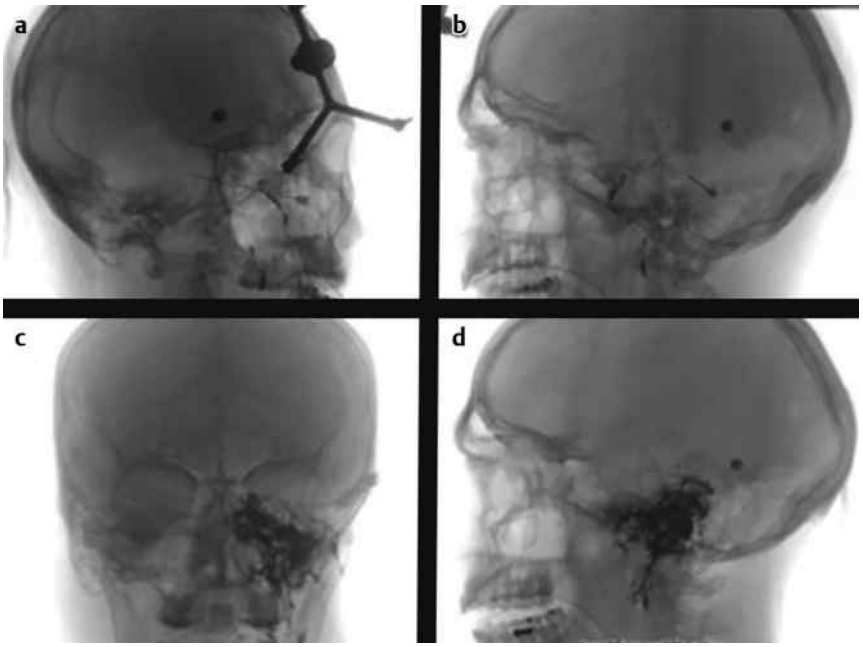

Fig. 21.13 Direction puncture embolization. (a) Oblique and **(b)** lateral views of the patient during direct-puncture embolization prior to injection of Onyx. Note the spinal needle placed into the tumor with navigation and fluoroscopy guidance. **(c)** Anteroposterior and **(d)** lateral views demonstrated embolization material within the tumor after direct-puncture embolization.

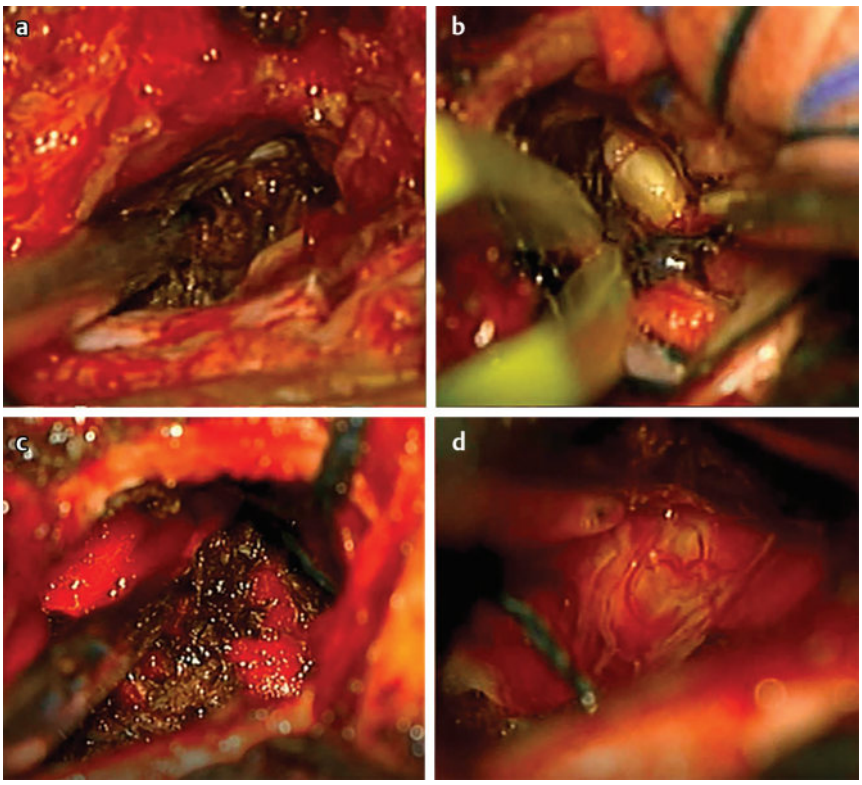

Fig. 21.14 Intraoperative images. **(a)** Initial tumor exposure after removal of previous mesh and dural opening; **(b)** subsequent dissection of the tumor along the cerebellar surface to the brainstem, where the brainstem/tumor interface was exposed. **(c)** After this plane was carefully developed, the tumor was partially debulked and freed from its petrous attachments. **(d)** View of the operative field after the posterior fossa tumor was removed.

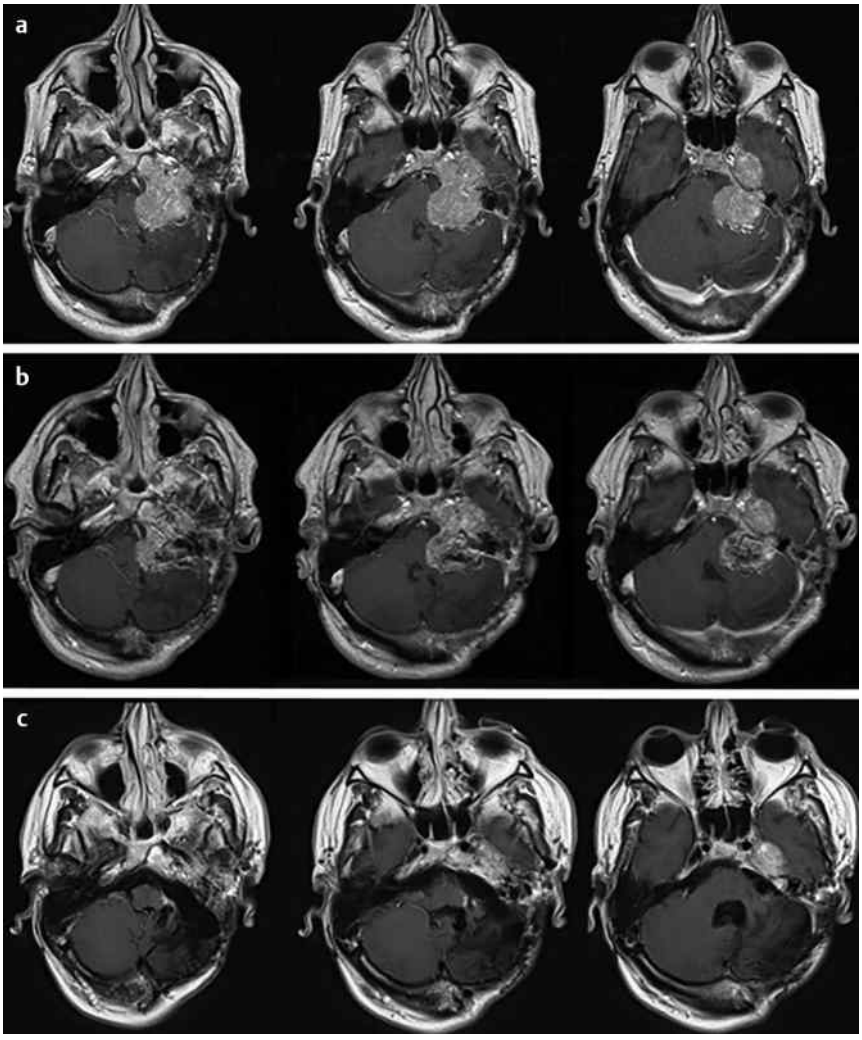

Fig. 21.15 Axial MRI at various stages of treatment. **(a)** MRI on presentation, before current series of treatment; **(b)** MRI taken after direct-puncture embolization of the intracranial portion of the tumor showing penetration of Onyx mainly into the posterior fossa component of the tumor; **(c)** postoperative MRI showing complete decompression of the brainstem, with remaining tumor in the cavernous sinus.

Surgical Pearls

1. For functional preservation and mitigation of surgical risk, especially bleeding, multimodality treatment is often necessary for recurrent, progressive glomus jugulare tumors. It is ideal for specialists to meet as a panel to coordinate plans and formulate a consensus strategy prior to commencement of any individual treatment arm.
2. Massive operations for aggressive tumor removal, which involve carotid artery bypass or iatrogenic destruction of intact function, may not serve the patient's best interest since glomus jugulare tumors evolve very slowly.
3. Direct-puncture embolization is an alternative for elimination of the tumor's blood supply when catheter access is not feasible. Although not as reliable as catheter embolization, it can still be quite effective.

Aftercare

Postoperatively, the patient did well except for a HB 6 facial nerve weakness. Aggressive eye care with taping and patching of the affected side prevented injury to the eye itself. The remainder of his neurological exam was unchanged after surgery. However, on postoperative day 4, clear drainage from his incision was noted. The incision was oversewed where a focal area of drainage was noted. A frontal external ventricular drain was placed to address the CSF leak and rule out an elevated intracranial pressure (ICP). Several days of continuous CSF divergence allowed the incision to heal and as the ICP was normal, the drain was removed.

Other potential complications of surgeries for glomus jugulare tumors include injuries to lower cranial nerves. The patients' ability to protect their airway must be carefully assessed before removal of the endotracheal tube and initiation of oral feedings. Fortunately, our patient had habituated to his previous lower cranial nerve palsies, and neither tracheostomy nor gastrostomy was necessary for him.

According to the plan laid out by the multidisciplinary panel, the patient underwent radiosurgery for this residual tumor approximately 6 months following surgery. The extracranial tumor in the infratemporal/pterygoid fossa and neck, as well as the remaining intracranial tumor in the cavernous sinus and middle fossa were treated with 25 Gy in 5 fractions (**Fig. 21.16**).

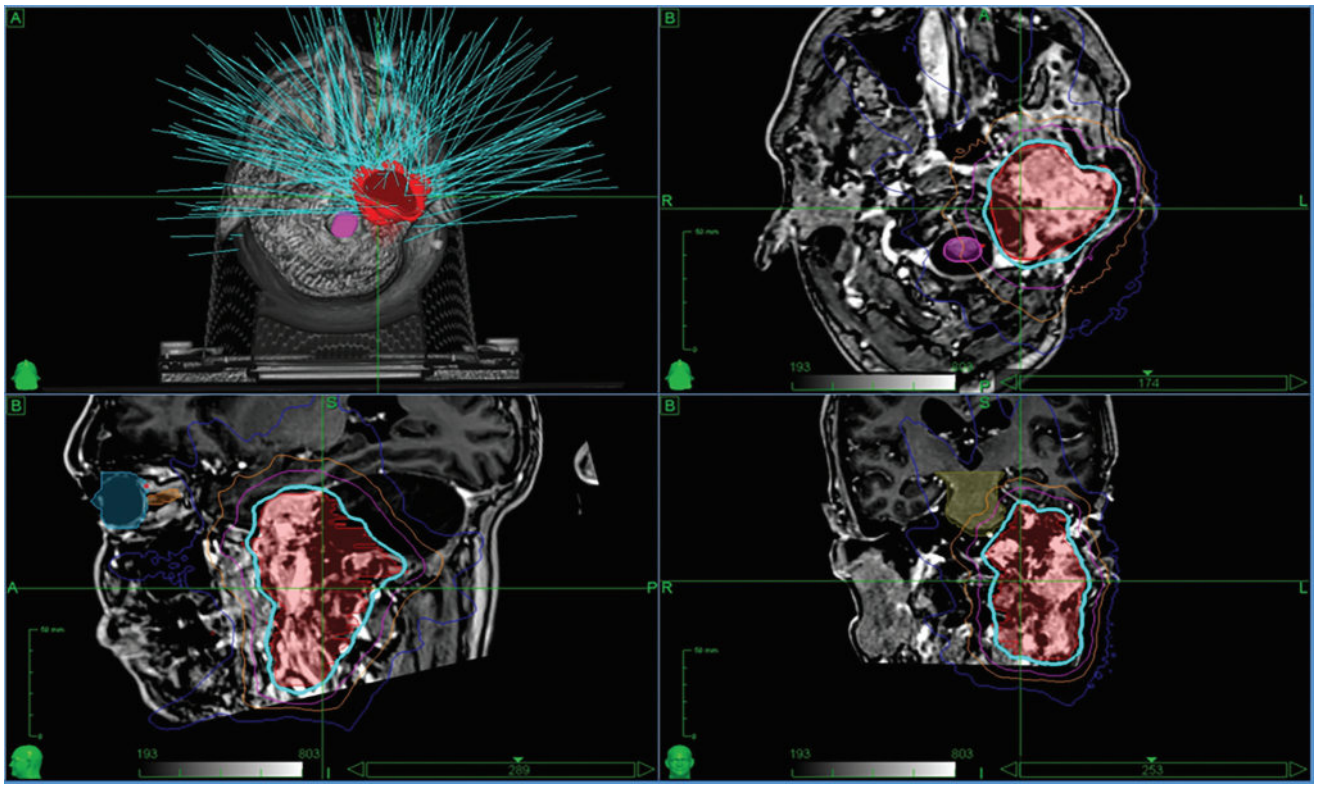

Fig. 21.16 Planning images for postoperative radiosurgery. The extracranial tumor was intentionally left for postoperative radiosurgery. The blue line indicated the margins of the radiosurgery treatment plan. The tumor within the blue line was treated to 25 Gy in 5 fractions.

Perspective

Luis A.B. Borba and Marcio S. Rassi

■ Introduction

Paragangliomas of the jugular foramen or glomus jugulare tumor are generally benign, slow-growing neoplasms, and their treatment has significantly changed over the past decades. For many years, these tumors were deemed inoperable because of their deep location, complex anatomical entanglement with neighboring structures and high risk of severe intraoperative bleeding. As such, radiation therapy became the first option for treatment. However, the analysis of tissues from irradiated tumors revealed that despite increased amounts of fibrous tissue, the tumor cells remained viable. This leads us to conclude that the efficacy of radiation therapy is only temporary.

■ Assessments

Factors such as patient's age and symptoms, as well as tumor size and secretory status, must be considered before choosing the proper treatment of glomus jugulare tumors. Small lesions (Fisch A and B) in older patients (over 65 years of age) that are asymptomatic or minimally symptomatic may be observed. In patients whose symptoms are progressive, radiosurgery for growth control can be offered. Young, symptomatic patients with preserved lower cranial nerve function are good candidates for surgical resection. Our experience has shown that in this situation, the anterior wall of the jugular bulb can be preserved to avoid impairment of lower cranial nerve function, and a curative surgical removal is possible. For large tumors (> 2 cm), we believe that radical surgical removal is the best choice of treatment.

The preoperative identification of a secretory tumor is mandatory for surgical planning. In addition, genetic studies should be performed to identify potentially malignant tumors. The tumor's blood supply must be carefully studied before surgery, but preoperative embolization is not suitable for all cases. We avoid this procedure in cases where the functions of the lower cranial nerves are preserved, because of potential risk to their blood supply. In cases with preexisting lower cranial nerve dysfunction, embolization is indicated; however, surgery must be performed within 48 hours of the endovascular procedure. Outside this time window, the blood supply of the tumor can shift from the external to internal carotid circulation, making resection more difficult.

■ Anatomical and Therapeutic Considerations

When surgical treatment is decided, the choice of the best surgical approach will be determined by several anatomic criteria. However, all the approaches have in common certain standardized steps in surgical technique, the anatomic basis of which is shown in **Fig. 21.17**.

1. Pericranium, fascia, and muscular flaps must be prepared for closure. The posterior half of temporalis muscle is rotated inferiorly, attached to the SCM muscle and its fascia, and is used, if necessary, to repair any dural defect.
2. The neurovascular structures of the neck are dissected; the posterior belly of digastric muscle is detached and displaced anteriorly.
3. The basic mastoidectomy is performed, exposing the sigmoid sinus from its junction with the transverse sinus to the jugular bulb. The IJV is dissected cranially until the jugular bulb is reached. The jugular foramen is opened from the mastoid cavity and inferiorly, exposing the whole length of the venous system, from IJV to the lateral segment of transverse sinus.
4. The ICA is isolated and followed cranially.
5. The ECA and its branches are identified and ligated and/or coagulated.
6. The sigmoid sinus is skeletonized and closed distally, just after the take-off of posterior emissary vein or as proximal as the extension of the tumor.
7. If the lower cranial nerve functions are preserved, generally the anterior wall of the jugular bulb is intact. Bleeding from this area is commonly from venous channels that can be occluded with fibrin glue or similar material. Excessive coagulation must be avoided to preserve the vascularization of the nerves, which runs in close relation to the anterior wall of the jugular vein.
8. Intradural extension can be removed by following the tumor with special care to preserve all neurological structures.

Options for Approach

The next steps of the procedure are tailored to each case and based on the preoperative symptoms, tumor extension and blood supply.

For tumors located in the jugular foramen, with or without neck or intradural extension, with no extension to the middle ear or surrounding the petrous ICA, to which the blood supply is essentially from branches of the *ECA*, we prefer to use the transmastoid, infralabyrinthine approach, which has been described in the previous section in this chapter. For this, the facial nerve is kept in its anatomical position, and the fallopian canal is untouched. The external auditory meatus is preserved, and the middle ear structures are kept intact. The tumor is removed from caudal to cranial, and neck/intradural extensions are followed and removed. Both facial nerve and hearing function can be preserved.

When the blood supply to the tumor is from the *ICA* and from tympanic branches, a complete control of the ICA is mandatory. If the tumor extension to the middle ear does not affect the anatomical structures, and the hearing is preserved, the approach must include the drilling of the tympanic bone and skeletonization of fallopian canal. This resembles the aforementioned "fallopian bridge" in the preceding section (**Fig. 21.18**). The tympanic membrane is then displaced superiorly. An infralabyrinthine route, pre- and retrofacial, is used. In the end, the tympanic membrane is reconstructed, and the external auditory canal is kept opened.

Some tumors have anterior and superior extensions, following and encasing the cervical and petrous ICA, and yet, the facial

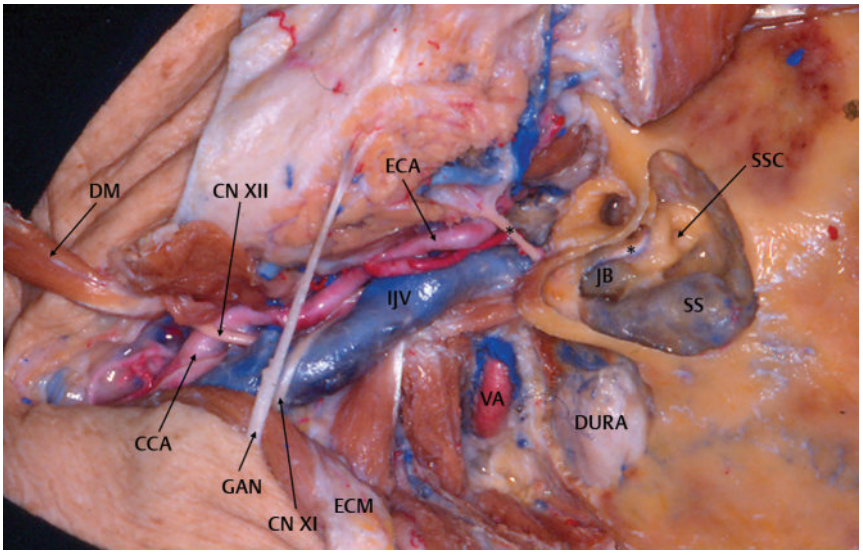

Fig. 21.17 Transmastoid infralabyrinthine approach. Left-sided cadaveric dissection showing the basic platform of the transmastoid, infralabyrinthine approach. CCA, common carotid artery; CN, cranial nerve; DM, digastric muscle; ECA, external carotid artery; ECM, sternocleidomastoid muscle; GAN, great auricular nerve; IJV, internal jugular vein; JB, jugular bulb; SS, sigmoid sinus; SSC, semicircular canals; VA, vertebral artery; asterisks, facial nerve.

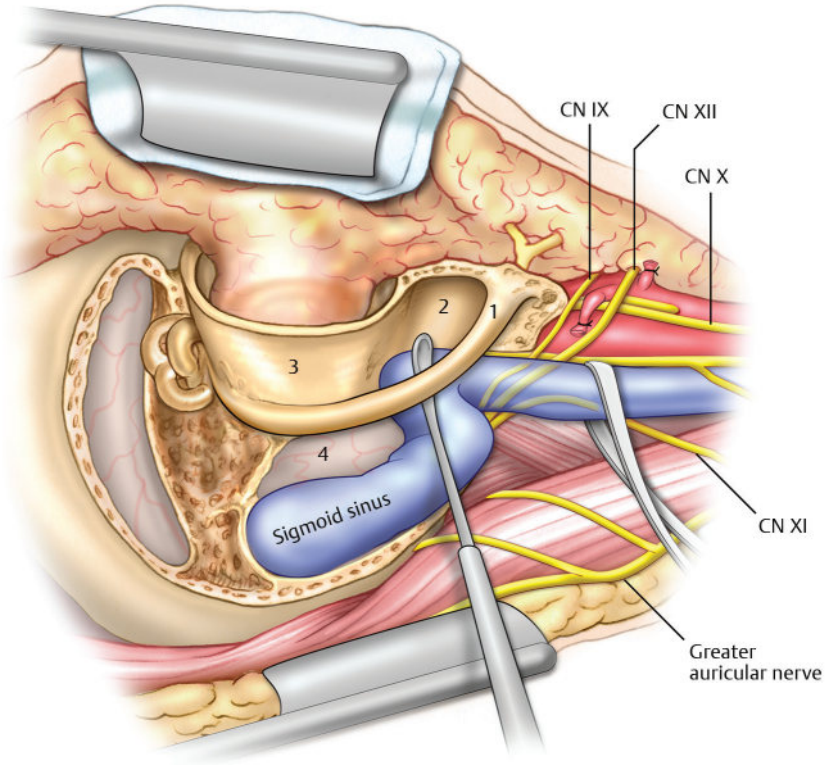

Fig. 21.18 Fallopian bridge. Illustration demonstrating the concept of the fallopian bridge. The facial nerve is "skeletonized" in its native bony canal, and the resection of the tumor takes place anterior and posterior to it. Please note that in this illustration, the canal wall and external acoustic meatus are kept intact. 1: the fallopian bridge; 2: bone anterior to the facial nerve, removed; 3: posterior canal wall; 4: posterior fossa dura.

nerve function remains intact. In such situations, the external auditory meatus is closed in sac, the middle ear structures removed, the oval window plugged, and the ICA is followed from the neck to its petrous and cavernous segments. The facial canal is skeletonized, but the facial nerve is kept in anatomical position. The tumor is removed anterior and posterior to the facial nerve, but special care must be taken with the geniculate ganglion, located near the floor of middle fossa, to avoid injury.

The tumor can grow to giant sizes, destroying all the temporal bone structures, facial nerve, hearing, and lower cranial nerve function. In this situation a total petrosectomy is planned, the ICA is followed cranially, and the facial nerve is identified and grafted with the greater auricular or sural nerve. The external auditory canal is closed in sac, the invaded dura mater repaired with dural graft, and the large cavity filled with abdominal fat.

Case Presentation 1

A 31-year-old man presented with a 6-month history of left-sided deafness, dysphagia, and hoarseness. His neurological examination revealed a left-sided atrophy of the tongue, loss of hearing and weakness in shoulder elevation. Indeed,

cranial nerves VIII through XI were dysfunctional on the left side. His MRI showed a paraganglioma involving the left jugular foramen and the digital angiography demonstrated a highly vascular tumor fed mainly by the ECA (**Fig. 21.19**). The tumor was removed via an infralabyrinthine approach (**Fig. 21.20**). The patient's postoperative course was unremarkable and he did not experience any new neurological deficits. The MRI after surgery showed a complete resection (**Fig. 21.21**).

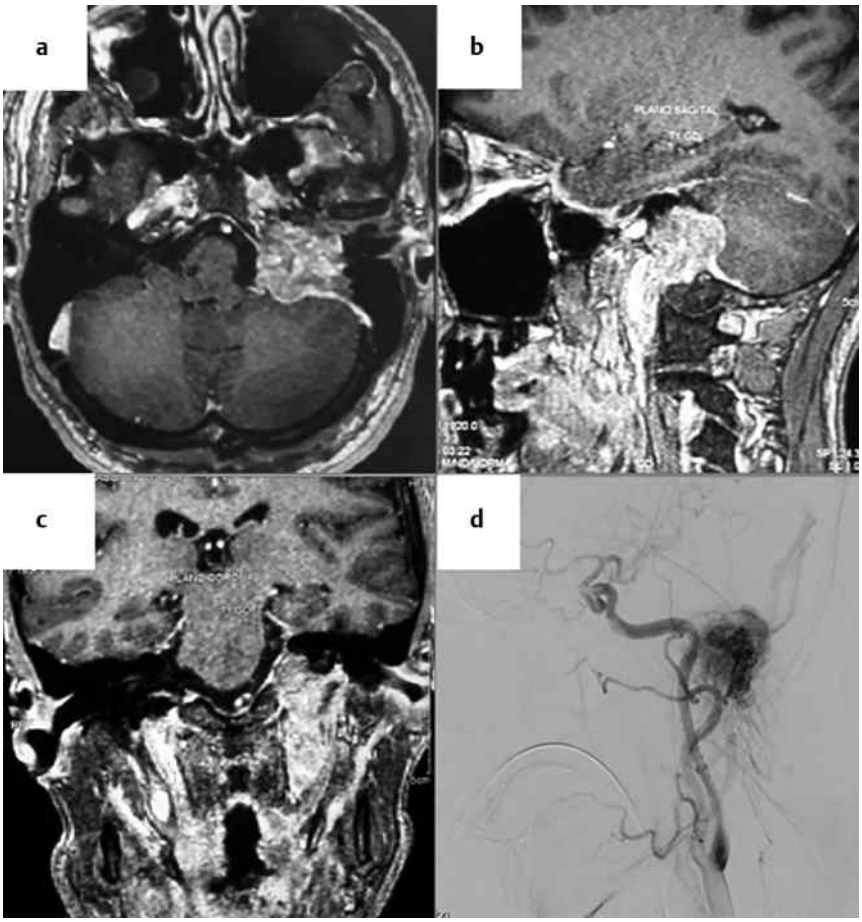

Fig. 21.19 Images of Case 1. Preoperative **(a)** axial, **(b)** sagittal, and **(c)** coronal post-contrast T1-weighted MRI demonstrating a lesion involving the jugular foramen suggesting a paraganglioma. **(d)** Digital angiography depicting a highly vascularized tumor fed mainly by the ECA.

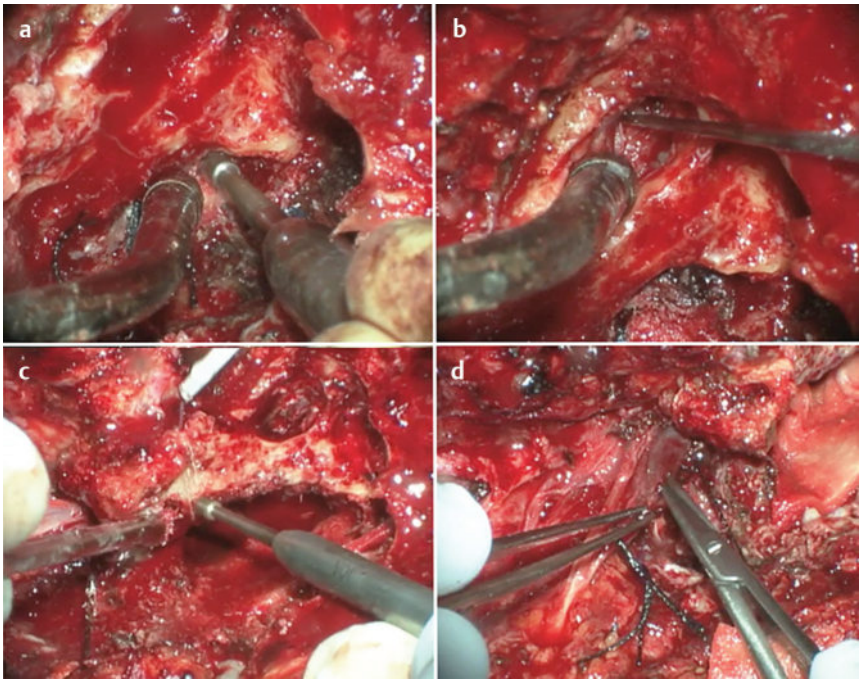

Fig. 21.20 Intraoperative images of Case 1. **(a)** Drilling inferior to the semicircular canals during the infralabyrinthine approach; **(b)** removing tumor from the middle ear; **(c)** drilling close to the facial nerve while leaving it unharmed; **(d)** opening the jugular bulb for tumor removal.

■ Management of Recurrent Tumors

The treatment of a recurrent glomus jugulare tumor is very difficult. In our practice, recurrence is considered if any tumor growth is noted between 6 and 12 months following treatment—excluding residual lesion identified in the immediate MRI. Patients with clearly identified recurrences without cranial nerves deficits would be offered radiosurgery. However, if the patient already has lower cranial nerve deficits, whether or not this is associated with new facial nerve palsy, we prefer surgical treatment for radical and potentially curative removal.

For a recurrent case, such as the one presented in the first section of this chapter, our management decision would have been heavily influenced by the patient's poor cranial nerve function, his previous surgical and radiation therapy, as well as the large size of the tumor. We would perform a preoperative

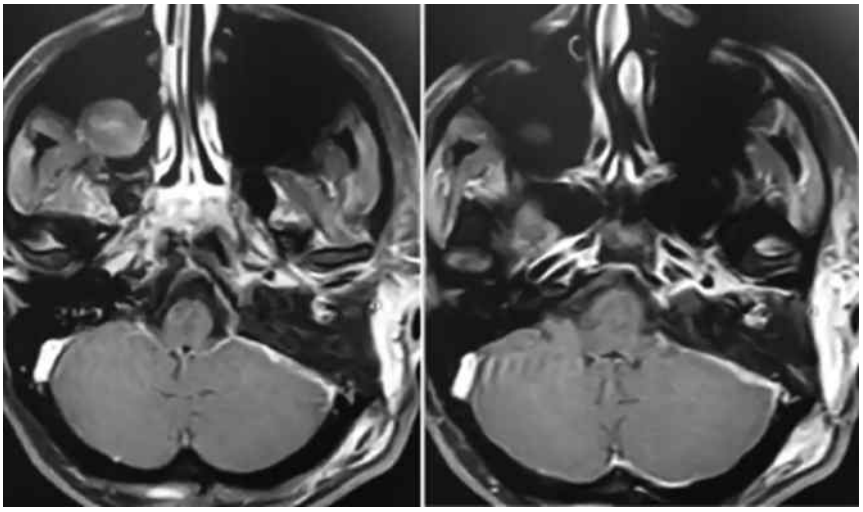

Fig. 21.21 Images of Case 1 after surgery. Axial postcontrast T1-weighted MRI showing complete tumor removal through an infralabyrinthine approach.

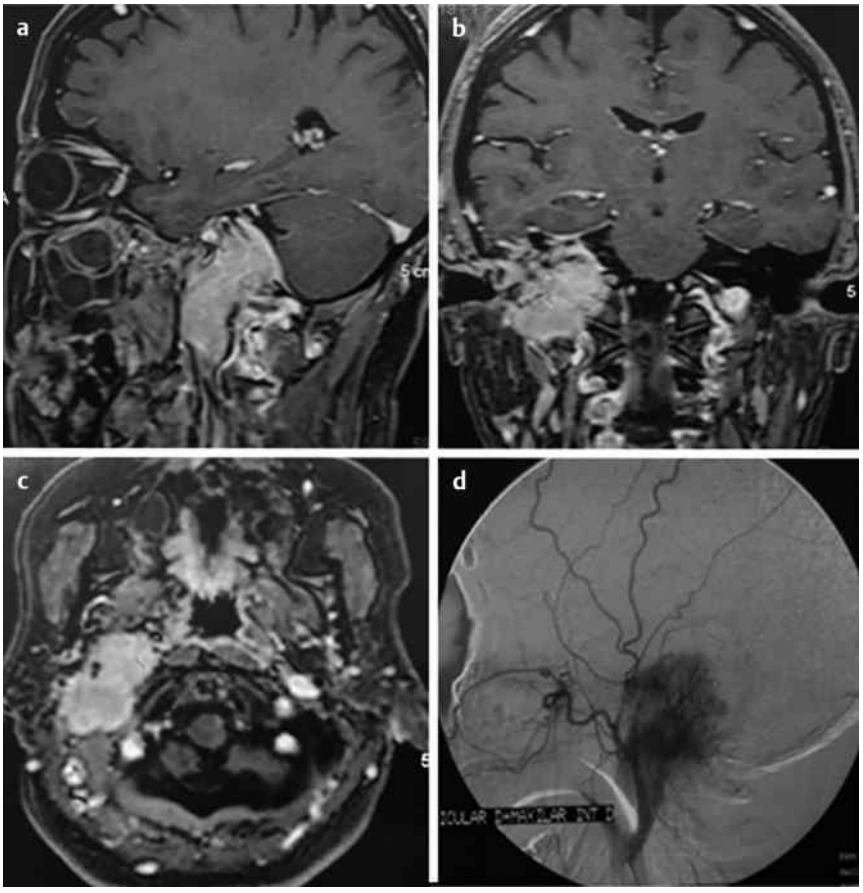

Fig. 21.22 Images of Case 2. **(a)** Sagittal, **(b)** coronal, and **(c)** axial MRI with gadolinium showing a recurrence of the patient's previously resected tumor. **(d)** Digital angiography depicting a lesion mainly vascularized by the ICA.

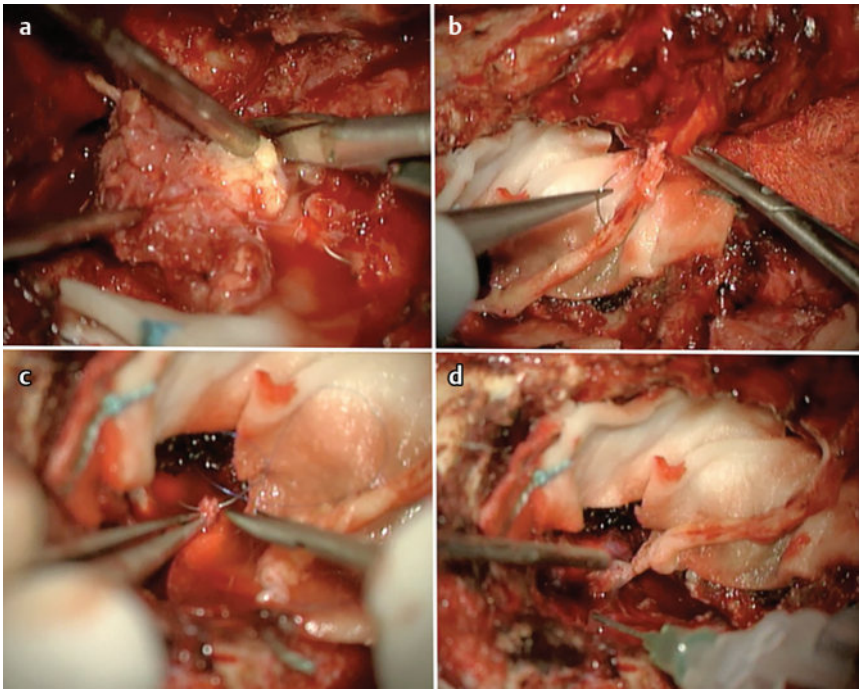

Fig. 21.23 Intraoperative images of Case 2. **(a)** The tumor was removed via a radical sided petrosectomy; **(b)** the sural nerve interposition graft was sutured to the distal stump of the facial nerve; **(c)** the same graft was sutured to the proximal stump at the cerebellopontine angle; **(d)** the full interposition graft in its final position. The patient's facial nerve function improved over the first postoperative year.

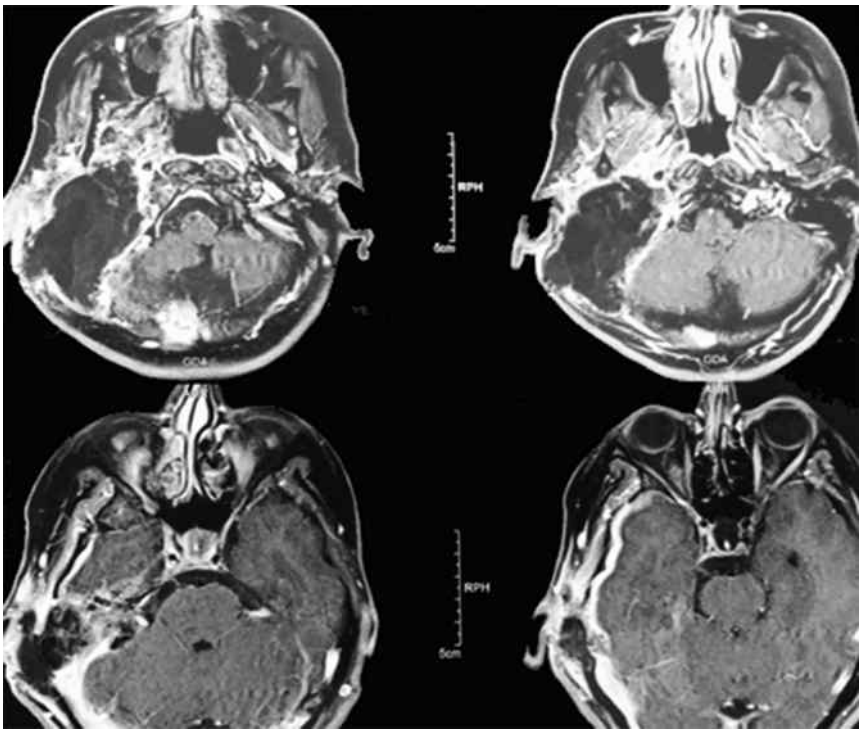

Fig. 21.24 Postoperative images of Case 2. Axial postcontrast T1-weighted MRI showing complete tumor removal through a total petrosectomy. The facial nerve was grafted using the sural nerve (the greater auricular nerve was not available due to the previous surgery).

balloon occlusion test to analyze the collateral blood supply, and if the patient passed the test, plan a radical total petrosectomy to aim for total removal of the tumor, in either a single- or a two-stage operation. If, however, the patient failed the balloon test occlusion, then we would plan for leaving tumor residual surrounding the ICA for further radiotherapy.

Case Presentation 2

A 32-year-old woman experienced a recurrent glomus jugulare tumor. She had previous surgery at another institution and presented to us with right-sided facial palsy, deafness, hoarseness, and mild dysphagia. Her MRI showed a recurrent tumor and the digital subtraction angiography revealed a blood supply mainly by the ICA (**Fig. 21.22**). A total petrosectomy was performed to remove her tumor. Since the greater auricular nerve was not available due to her previous operation, we used the sural nerve as an interposition graft to reconstruct her facial nerve from the CPA to the parotid segment (**Fig. 21.23**, **Video 21.2**). The skull base defect was closed with fascia lata, muscle rotation, and abdominal fat graft. After surgery, the MRI showed a complete resection (**Fig. 21.24**). She had no new deficits, and her facial nerve function improved significantly after 1 year.

Commentary

The only curative treatment for glomus jugulare tumors is radical total removal. We must balance the risks and benefits of it, because radiation therapy can offer acceptable long-term disease control in some cases. Preoperative symptoms, tumor extension, clinical condition, and genetic factors should be considered in the decision-making process. Despite the apparently complex surgical technique, a detailed understanding of regional anatomy, careful intraoperative technique, and meticulous postoperative care can contribute to successful surgical treatment, leading to a cure for the patient with preservation of cranial nerve functions.

Recommended Reading

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Part VII

Tumors of the Posterosuperior Skull Base

22 Falcotentorial *285*

23 Superior Vermis *298*

24 Pineal *308*

22 Falcotentorial

Hussam Abou-Al-Shaar, Neil Majmundar, and James K. Liu

Keywords: vein of Galen, falx cerebri, corpus callosum, tentorium, transverse sinus reanastomosis

■ Case Presentation

A 60-year-old man presented to our service with two episodes of complete visual loss, each lasting 10 minutes. He also complained of headaches, worsening balance, dizziness, memory difficulty, and left upper extremity weakness and paresthesias. The patient has a history of a pineal region, falcotentorial mass that was observed for the past 10 years. His neurological examination was unremarkable. Magnetic resonance imaging (MRI) depicted significant increase in the size of the mass from 3.2 to 4.7 cm (**Fig. 22.1**). The patient was admitted to the hospital for further investigations and management.

Questions

1. What are the common signs and symptoms for lesions in this location?
2. How does the differential diagnosis influence the next step in management? What further tests should you consider?

■ Diagnosis and Assessment

The MRI demonstrated a homogenously enhancing pineal region falcotentorial mass. The differential diagnosis for the tumor includes meningiomas, germ cell tumors, pineal gland tumors, gliomas, metastasis, and vascular malformations.

Patients with tumors located in the pineal region can present with a variety of signs and symptoms depending on the size of the tumor and its relationship with adjacent structures. Headache is the most common complaint. Others include dizziness, gait ataxia, papilledema, memory difficulties, hearing impairment, visual changes, weakness, paresthesias, urinary incontinence, and signs of increased intracranial pressure. Moreover, many of these patients present with Parinaud's syndrome, but the incidence of this among patients with meningiomas is lower compared to patients with other pineal region tumors. One possible explanation for this is that falcotentorial meningiomas grow slowly in the cistern, and cause less disturbance to the periaqueductal region than pineal tumors. However, many of these meningiomas progress to obstruct the aqueduct and eventually cause symptomatic hydrocephalus.

It is essential to measure tumor markers to rule out germ cells tumors, which are better addressed with nonsurgical options including chemotherapy and/or radiation therapy. However, tumor markers in our patient were negative. With this, and taking into account the location, degree of enhancement, presence of a dural tail, and the broad-based attachment to the falcotentorial dura, a meningioma was the most likely diagnosis in our case.

When a pineal region mass is considered for surgical intervention, it is important to obtain further imaging to understand the tumor's relationship to adjacent vascular structures, and determine the degree of venous involvement within the tumor. Thus, a computed tomography angiogram/venogram (CTA/CTV), MR angiogram/venogram (MRA/MRV), or a conventional angiogram should be obtained in preparation for definite surgical intervention. Conventional angiogram would permit further analysis of the feeding vessels and possible preoperative embolization of the tumor. In our patient, the tumor was situated superior and dorsal to internal cerebral veins and vein of Galen, with significant mass effect on the splenium of the corpus callosum and lateral displacement of the parietal lobes (**Fig. 22.2**). Given the patient's clinical deterioration and the tumor growth seen on this MRI, the decision was made to proceed with surgical removal, with the aim of gross total resection (Simpson grade I).

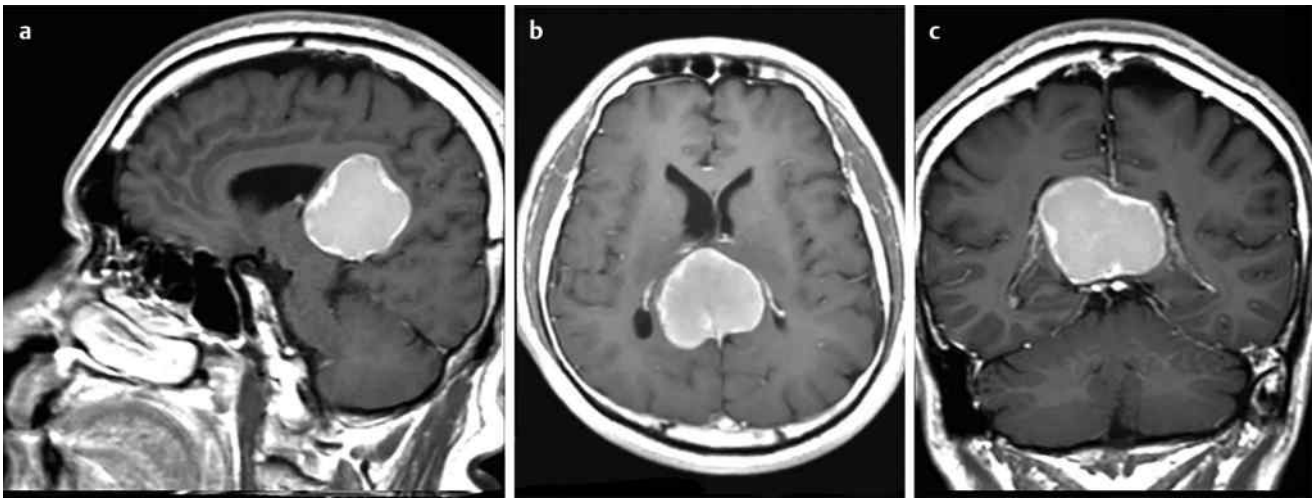


Fig. 22.1 Preoperative post-gadolinium T1-weighted MRI. (a) Sagittal, **(b)** axial, and **(c)** coronal view demonstrated a large falcotentorial meningioma located in the pineal region.

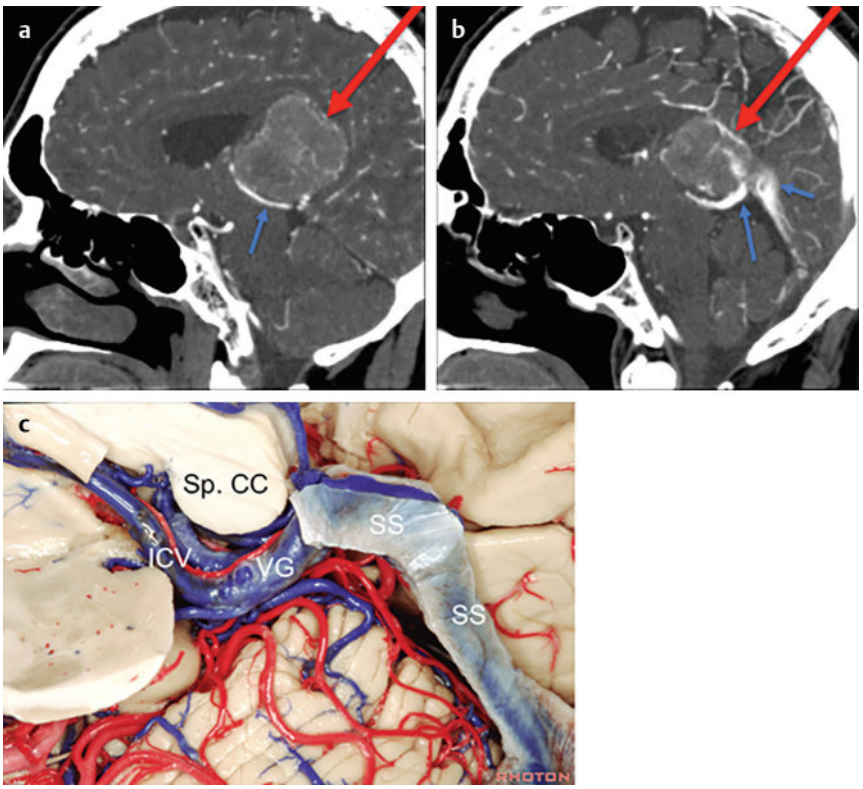


Fig. 22.2 Preoperative assessment. CT angiogram (sagittal view) **(a)** and **(b)** showed the location of the tumor superior to the vein of Galen complex and internal cerebral veins (*blue arrows*). The trajectory to the tumor was a posterior interhemispheric retrocallosal approach (*red arrow*). **(c)** Sagittal view of the splenium of the corpus callosum (Sp. CC), internal cerebral veins (ICV), vein of Galen (VG) and straight sinus (SS). (The Rhoton Collection.)

■ Anatomical and Therapeutic Considerations

Falcotentorial meningiomas arise from the junction of the dura of the tentorium cerebelli and posterior part of the falx cerebri. These tumors are extremely rare, accounting for 0.3 to 1.1% of all intracranial meningiomas. The surgical approach for these lesions depends on the location of the tumor, laterality, relationship to adjacent neurovascular structures (especially the internal cerebral vein and vein of Galen), involvement/patency of adjacent venous structures, and the development of collaterals (**Fig. 22.3**). Taking these into account, the ideal approach should additionally provide the most direct route and trajectory to the tumor without jeopardizing the adjacent venous channels.

Because of this complexity, various authors have classified falcotentorial meningiomas into different groups, based on their relationship and displacement of the adjacent vascular structures, to optimize selection of the surgical approach. The major vascular supply of falcotentorial meningiomas comes from the tentorial branches of the meningohypophyseal trunk, the artery of Bernasconi and Cassinari. Other possible feeders include the superior cerebellar artery, the posterior cerebral artery (PCA), posterior choroidal arteries, vertebral artery, and branches from the external carotid artery. In contrast, meningiomas arising from the velum interpositum get their vascular supply from the posterior choroidal arteries. It has been noted that falcotentorial meningiomas tend to displace the PCA laterally in the quadrigeminal cistern, and the posterior choroidal arteries are generally displaced anteroinferiorly, along with the internal cerebral veins and vein of Galen. In our patient, the venous complex was displaced inferiorly (**Fig. 22.2**). Of note, the absence of angiographic visualization of any venous channel due to tumor encasement does not necessarily mean that the vein can be sacrificed. That vein may yet be patent, and though the flow is minimal, it may be a critical part of a collateral network for venous drainage.

Options for Approach

Because of their scarcity, location, and presence of adjacent eloquent neurovascular structure, falcotentorial meningiomas represent a management challenge for neurosurgeons. Approach options include the occipital transtentorial, posterior interhemispheric retrocallosal transfalcine, supracerebellar infratentorial, bioccipital, and combined supra- and infratentorial approaches (**Fig. 22.4**). Optimal selection of the surgical access and working corridor is of paramount importance. Generally, falcotentorial tumors arising above the internal cerebral veins and vein of Galen are approached with the occipital transtentorial approach or, as in our case, with a posterior interhemispheric retrocallosal transfalcine approach (**Fig. 22.2**). On the other hand, the tumors that arise inferiorly to the internal cerebral veins and vein of Galen can be addressed utilizing the supracerebellar infratentorial approach. A combined supra- and infratentorial approach is reserved for large lesions extending in the sagittal/coronal planes, while the bioccipital approach is

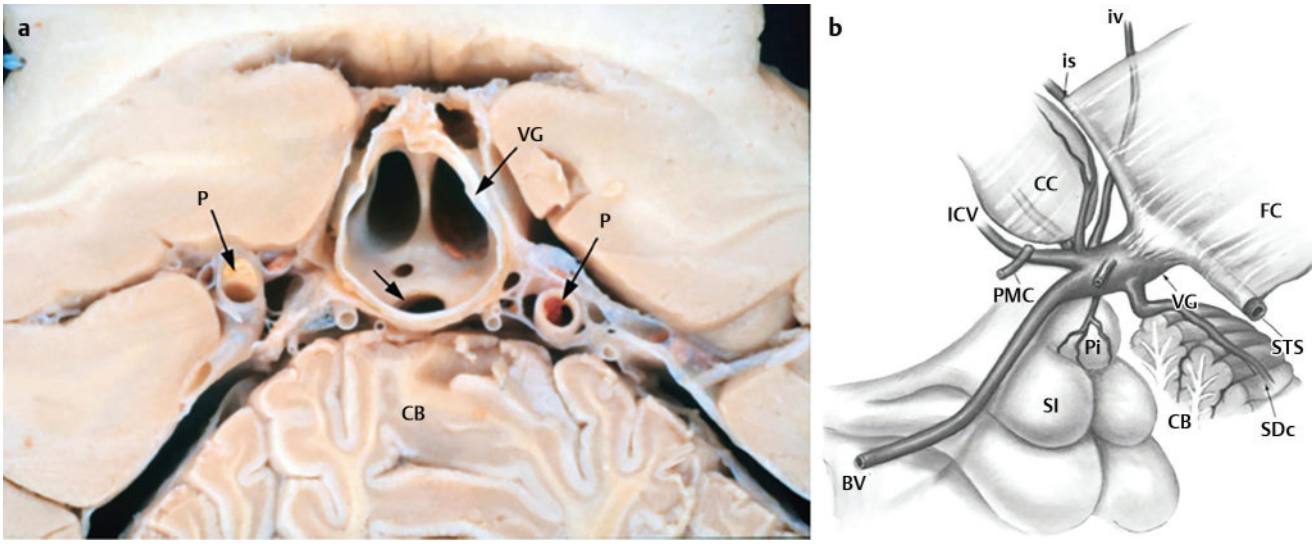


Fig. 22.3 Anatomy of the falcotentorial region. (a) A coronal section through the vein of Galen at the level of the splenium of the corpus callosum. This is just prior to the vein of Galen turning superiorly to enter the straight sinus. Arrow indicates the entrance of the superior cerebellar vein. Note the septum dividing the vein of Galen on either side of the entrances of the internal cerebral veins. **(b)** Illustration of a posterior and lateral view of the vein of Galen from the left side. BV, basal vein of Rosenthal; CB, cerebellum; CC, corpus collosum; FC, falx; ICV, internal cerebral veins; is, inferior sagittal sinus; iv, inferior cerebral vein; P, posterior cerebral artery; Pi, pineal; PMC, posterior medial choroidal artery; SDc, superior cerebellar vein; SI, superior colliculus; STS, straight sinus; VG, vein of Galen. (Reproduced from Grand W, Hopkins L, Siddiqui A, et al, eds. Vasculature of the Brain and Cranial Base. Variations in Clinical Anatomy. 2nd ed. Thieme; 2015.)

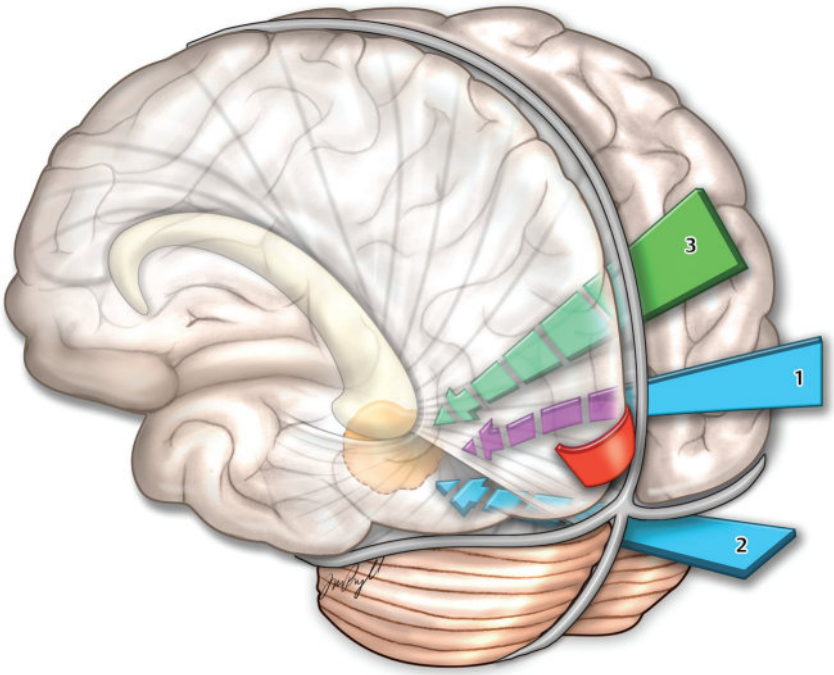


Fig. 22.4 Options for approach. The surgical approaches to a falcotentorial meningioma include the occipital transtentorial (*arrow 1*) and supracerebellar-infratentorial approach (*arrow 2*). The occipital approach can be performed bilaterally to make a bioccipital approach (*arrow 1 and red tail*). Alternatively, it can be combined with the supracerebellar-infratentorial approach (*arrows 1 and 2*) to make a combined supra- and infratentorial approach. The interhemispheric retrocallosal transfalcine approach (*arrow 3*) was chosen for the tumor in our patient.

reserved for large lesions extending laterally on both sides of the falx cerebri.

The occipital transtentorial approach is performed for superiorly arising falcotentorial tumors and is the most widely utilized approach for such lesions. It is generally performed on the right side with the patient in prone, sitting, or park-bench position. The key advantage of this approach is its spacious operative view to the pineal falcotentorial region, internal cerebral veins, and posterior and lateral midbrain. Moreover, if the tumor is extending to the contralateral side, an incision in the posterior falx (transfalcine approach) can be performed to access such lesions. The major disadvantage of this approach

is the risk of contralateral homonymous hemianopsia, which is primarily due to prolonged retraction of the occipital lobe.

Similarly, the posterior interhemispheric retrocallosal transfalcine approach offers great visualization of superiorly arising tumors that displace the deep venous system inferiorly. There is no risk of disconnection syndrome since the corpus callosum is left intact with this approach. The trajectory starts from the parieto-occipital region, and there is no retraction or compression to the occipital lobe. The view to the pineal region and falcotentorial junction is also more direct, and access to the contralateral side can be obtained through the transfalcine corridor.

The supracerebellar infratentorial approach is performed in the sitting position to allow the cerebellum to fall inferiorly; thus, the risk of air embolism is higher than other approaches. The major strength of this approach is with tumors arising from underneath the tentorium, near the junction of the vein of Galen with the straight sinus, as the veins would be displaced superiorly. Thus, the veins lie above the surgeon's working corridor between the tentorial edge and quadrigeminal plate. However, as the steepness of the tentorium increases, the approach becomes more challenging to perform.

The combined supra- and infratentorial approach is best suited for large lesions extending both superiorly and inferiorly that cannot be addressed using either the occipital transtentorial or infratentorial supracerebellar approaches alone. In this approach, the nondominant transverse sinus is usually sacrificed to enable greater operative exposure to the tumor. Despite the safety reports of this technique, many surgeons have encountered occipito-cerebellar hemorrhage following nondominant transverse sinus sacrifice, and some have recommended reconstructing the sinus after tumor resection to avoid this problem.

The bioccipital approach in the sea lion position is utilized for falcotentorial tumors extending laterally across the axial plane and having extensive adhesions to the surrounding vasculature. This approach allows for better exposure of the contralateral quadrigeminal region, as well as the supra- and infratentorial compartments, making complete removal of lesions extending bilaterally feasible. However, as in the combined supra- and infratentorial approach, the nondominant transverse sinus is sacrificed in this approach. Additionally, almost all patients develop transient cortical blindness after the bioccipital approach.

Regardless of the approach, if the tumor invades the surrounding vasculature and no dissection plane can be established between the tumor and the involved vessel, it is better to leave a residual tumor without harming the vessel. Specifically, the internal cerebral veins, straight sinus, and vein of Galen complex must be preserved at all costs, as injury to those vessels can result in devastating neurological injuries.

Approach of Choice

In our patient, the falcotentorial lesion was positioned superior to the internal cerebral veins and vein of Galen complex. Therefore, we elected to proceed with the posterior parietal interhemispheric retrocallosal approach because it provided the most direct access to the lesion with less risk of injury to the deep venous system (**Fig. 22.2**). We planned to utilize the endoscope to assist removal of the tumor, as it offered the dual-advantage of bringing illumination into a deep operative field and the ability to look behind "hidden" corners. Since preoperative imaging revealed patency of the straight sinus, meticulous dissection and preservation of the deep venous structures and collateral channels was of paramount importance.

Questions

1. What are the indications for the posterior interhemispheric retrocallosal transfalcine approach?
2. What are the technical nuances for the posterior interhemispheric retrocallosal transfalcine approach?
3. What are the expected outcomes and potential complications during falcotentorial tumor surgery?

■ Description of the Technique

The patient was positioned lateral with left side down to allow gravity-assisted displacement of the left parietal lobe away from the falx cerebri, as well as to bring the tumor mass on the right side into the operative field. The head was affixed to the Mayfield head holder, and positioned so that the falx was almost 30 degrees from the horizontal plane, which facilitates gravity-assisted access to the interhemispheric fissure without the need for fixed retraction on the dominant parietal lobe. Neurophysiological monitoring is of a great value in these cases and should be utilized (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: parieto-occipital interhemispheric, retrocallosal

Craniotomy: left parietal

Modifier: incision of falx

We elected to enter from the left side since the tumor extension was more lateral toward the right. Using a linear bicoronal incision, a left sided craniotomy was performed that crossed the midline to the right side. This allowed us to incise the dura on the left in a "C"-shaped fashion and reflect the dura toward the superior sagittal sinus to facilitate access to the interhemispheric fissure. A retractor was used to elevate the superior sagittal sinus and expose the hypervascular falx and tumor on the left side. Bipolar cautery was used to devascularize the hypervascular falx and tumor. Without the need of a fixed retractor on the left parietal lobe, the tumor on the left side was exposed and visualized (**Fig. 22.5**). Tumor debulking was then performed with the aid of an ultrasonic aspirator followed by extracapsular dissection of the tumor from the adherent left parietal lobe (see **Operative Setup**).

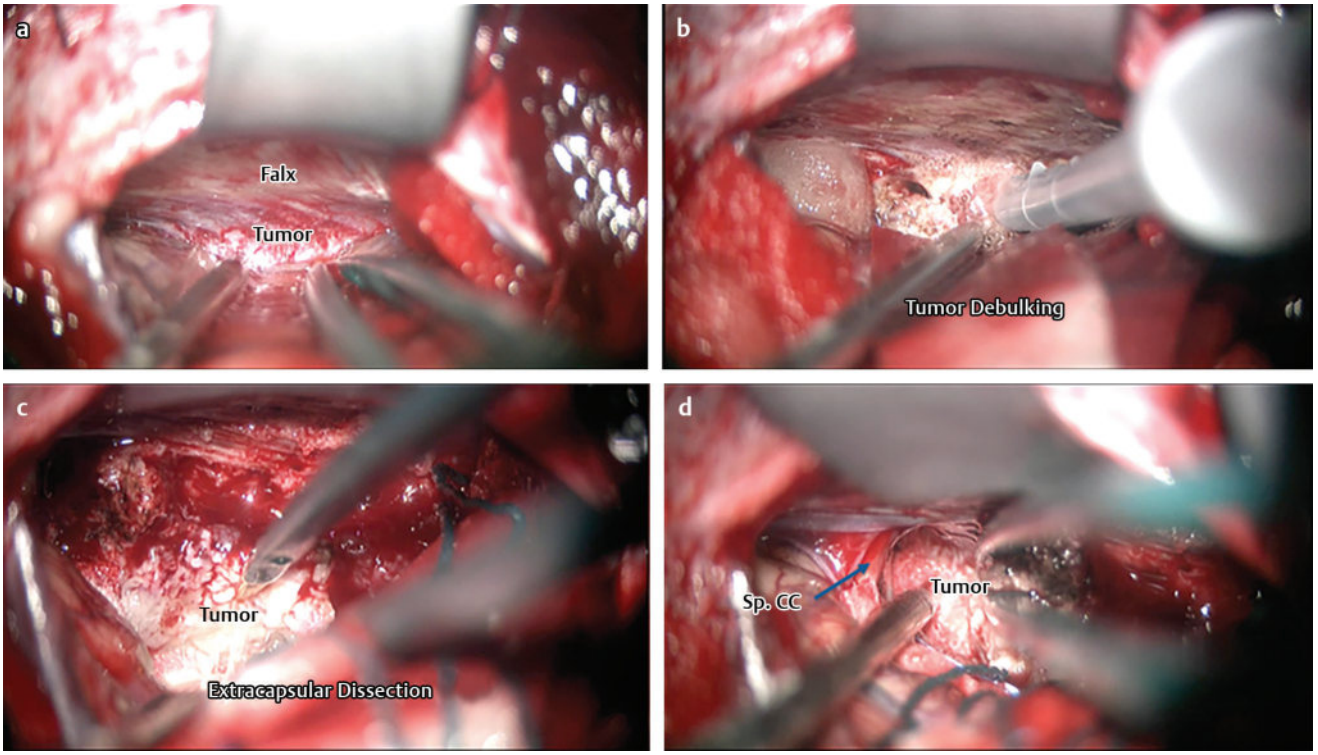


Fig. 22.5 Intraoperative images. (a) Interhemispheric view of the falx cerebri and tumor on the left side. **(b)** Tumor debulking was performed with an ultrasonic aspirator. **(c)** Extracapsular dissection was performed with bipolar forceps. **(d)** The tumor was dissected away from the splenium of the corpus callosum (Sp. CC) anteriorly.

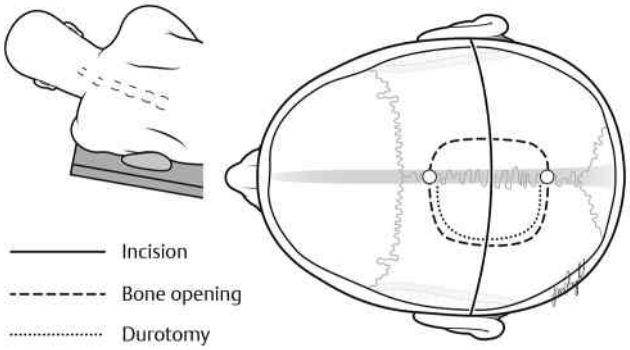
Operative Setup

Position: lateral, falx 30 degrees from horizontal

Incision: bicoronal

Bone Opening: burr holes over the SSS, left parietal craniotomy extending across midline

Durotomy: "C" shape reflected toward the midline/SSS



The splenium of the corpus callosum was visualized anteriorly and the retrocallosal corridor was used as the anterior plane for tumor dissection. The falx cerebri on the left side was incised to expose the tumor on the contralateral side (transfalcine approach) from the ipsilateral corridor (**Fig. 22.6**). Similarly, tumor dissection and debulking were carried out on the contralateral side. The tumor was removed in a piecemeal fashion. As dissection ensued along the posterolateral planes, the vein of Galen and its junction with the straight sinus were identified and preserved.

Following the removal of the tumor using the microscope, a 30-degree endoscope was brought to the operative field for inspection of the surgical cavity and corners that were not adequately visualized with the microscope (**Fig. 22.7**). The closure was performed in the standard fashion. Postoperative MRI demonstrated an excellent extent of resection with preservation of the deep venous structures and a small residual tumor adherent along the vein of Galen–straight sinus junction (**Fig. 22.8**).

Surgical Pearls

1. Patient positioning is the key for success. Optimal patient positioning in the lateral position will allow for gravity-assisted displacement of the ipsilateral parietal lobe and exposure of the ipsilateral tumor. Thus, rigid retraction on the ipsilateral parietal lobe is not needed.
2. Only with meticulous dissection of the tumor planes can the vein of Galen, straight sinus, and the internal cerebral veins be adequately visualized and protected. Moreover, dissection of the corpus callosum is also important because once the splenium is visualized, it demarcates the anterior plane of the tumor. From here, the retrocallosal corridor is sufficiently wide to remove the entire tumor, and there is no need to traverse the corpus callosum.
3. An angled endoscope at the end of microscopic tumor resection gives the surgeon better illumination and the ability to examine areas not sufficiently visualized with the microscope.

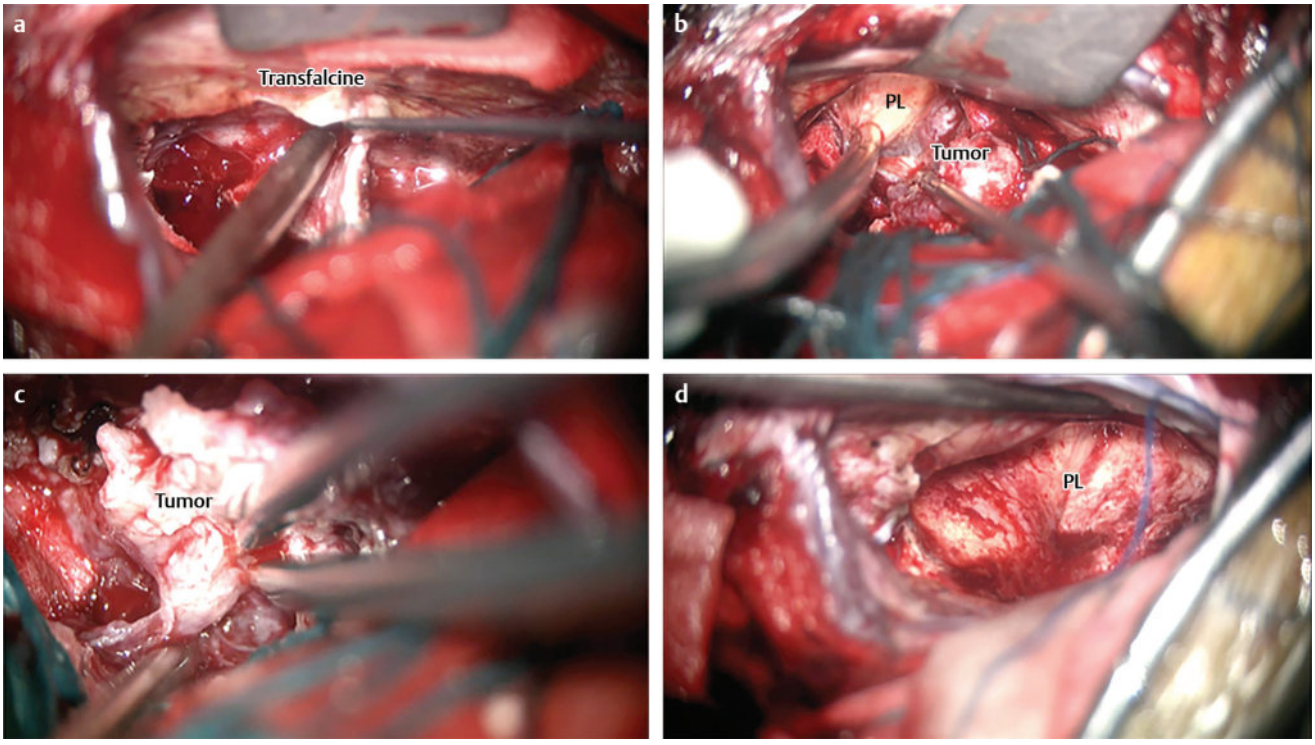


Fig. 22.6 Intraoperative images 2. (a) The falx cerebri was incised (transfalcine approach) to access the tumor on the right side of the falx. **(b)** The right side of the tumor was dissected away from the right parietal lobe (PL). **(c)** The tumor was delivered from the contralateral side. **(d)** The view of the resection bed of the right parietal lobe (PL).

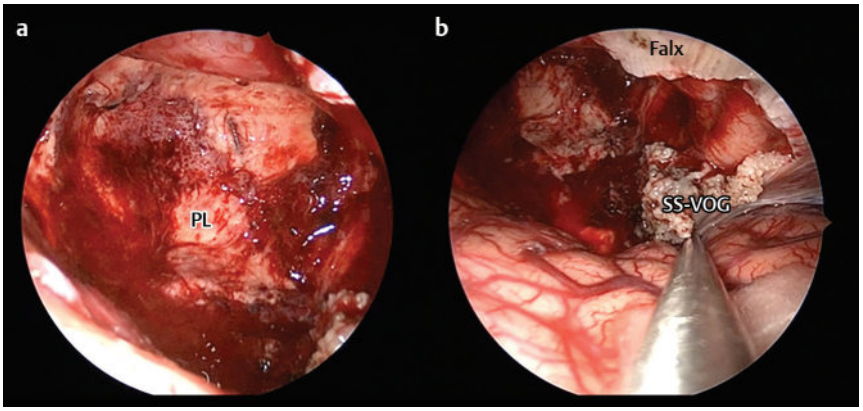


Fig. 22.7 Endoscopic views. (a, b) A 30-degree endoscope was used to inspect the resection cavity. The parietal lobe (PL) resection cavity was visualized and there was microscopic residual tumor invading the junction of the straight sinus and vein of Galen (SS–VOG).

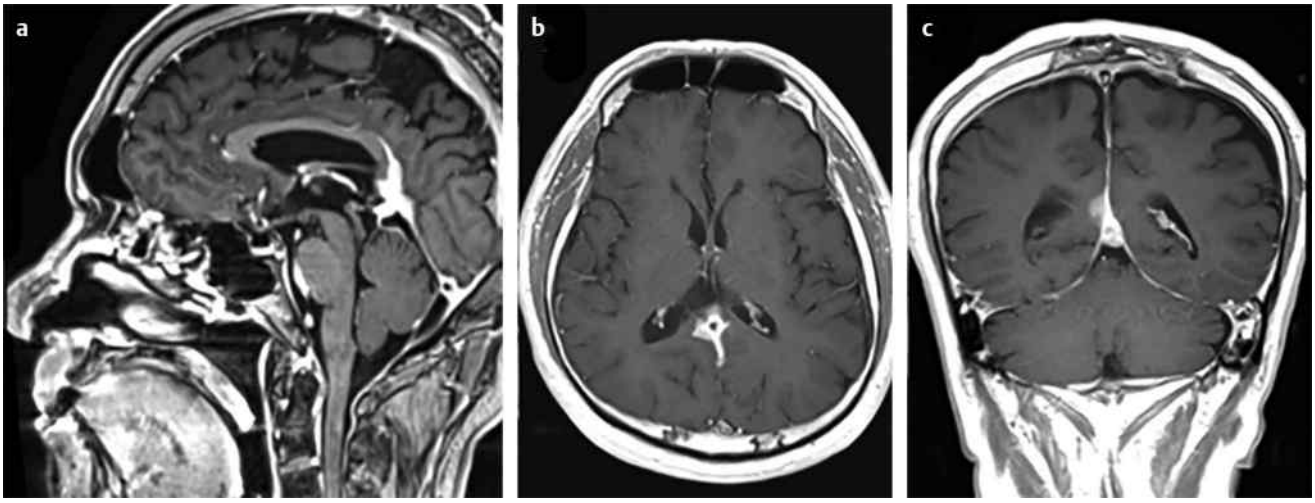


Fig. 22.8 Postoperative post-gadolium T1-weighted MRI. (a) Sagittal, **(b)** axial, and **(c)** coronal view showed a near-complete removal with a small residual tumor adherent to the vein of Galen complex.

■ Aftercare

The patient tolerated the procedure well and was discharged home with no neurological deficits. As mentioned earlier, postoperative MRI documented excellent resection with a small residual along the vein of Galen–straight sinus junction. Histopathological examination of the resected lesion depicted a WHO grade I meningioma. Since gross total removal of the tumor was not amenable in this case, the patient underwent stereotactic radiosurgery for the residual tumor.

■ Possible Complications and Associated Management

Complete surgical removal of falcotentorial meningiomas is reported in 50 to 100% of cases in various series. Residual and recurrent tumors respond well to radiation therapy. However, despite the high success rates for surgical resection of falcotentorial tumors, the morbidity rate is high, up to 23%. Various complications have been reported in the literature including homonymous hemianopsia, metamorphopsia, upward gaze palsy (cranial nerve IV palsy), hemiparesis, memory deficits, cerebrospinal fluid leakage, air embolism, brainstem ischemia, and intraventricular hemorrhage. Fortunately, the majority of these complications are transient. Meticulous dissection and preservation of the deep venous structures are essential in these cases. Injury to the deep venous system can lead to a myriad of severe neurological deficits too numerous to list, and therefore, preserving the deep venous structures should be the goal of surgery even at the expense of achieving a total resection of the tumor.

Perspective

Carolina Benjamin and Chandranath Sen

Introduction

Falcotentorial meningiomas are rare tumors that pose a surgical challenge given their deep location and close proximity to vital venous structures. When surgery is necessary, there are several considerations for selecting the ideal approach. These include: the best access across sensitive areas of the brain such as the occipital lobe, parietal lobe, and the cerebellum and how to minimize damage to them; optimal patient positioning; accessibility of the entire tumor and the related structures; protection of the venous sinuses, cortical draining veins, and the deep venous system; and finally, protection of the PCAs and their branches. The approach options to the falcotentorial region were mentioned previously in this chapter. The factors that influence the final selection can be divided into patient- and surgeon-related factors. The patient factors are dictated by the tumor, its surroundings, and individual variability of the venous anatomy, while the surgeon factors are related to the familiarity of a surgeon to the different approaches. Since these are uncommon tumors, a surgeon may encounter them but rarely, and that may influence the surgeon's choice of approach.

The authors of the previous section described a case where the posterior parietal interhemispheric retrocallosal approach was taken to remove a large falcotentorial meningioma. This tumor extended bilaterally, though it was slightly eccentric to the right. One of the benefits of their left-sided approach was that it provided better visualization of the contralateral portion of the tumor. Another benefit of this approach was that since the veins were displaced inferiorly by the tumor, the surgeons were able to debulk a majority of the tumor safely and create a larger working space for dissection of the deep venous complex during the late-phase of the operation.

While the splenium of the corpus callosum was pushed anteriorly by the tumor, there was still some tumor tucked anterior and inferior to the splenium that may have been difficult to access without splitting the splenium and risking a disconnection syndrome (**Fig. 22.9**). This may be one of the areas that required the usage of the 30-degree endoscope to look around "hidden corners" and improve the resection. While this approach worked very well for this tumor, as evidenced by the postoperative images and excellent clinical outcome, we would have chosen the occipital transtentorial approach.

The occipital transtentorial approach, just like the parietal approach, offers a direct and wide corridor to the tumor, but additionally, provides better visualization under the corpus callosum. Another benefit of this approach in comparison to the parietal interhemispheric retrocallosal approach is the relative paucity of cortical draining veins in the occipital region, when compared to the parietal. A wide surgical corridor is obtained with less risk to cortical veins, which decreases the risk of venous infarction. Another benefit of the occipital transtentorial approach is that it can be done in the prone position, with the chin tucked and the head elevated above the heart. This avoids complications such as herniation of occipital lobe out of the craniotomy defect, which can occur in the lateral position (and result in a hemianopsia). Finally, the surgeon can angle the microscope in all directions and rotate the table in all four planes to visualize tumor in deep and "hidden" areas without contorting into uncomfortable positions for prolonged periods. The main disadvantage of a prone position is that blood tends to pool at the bottom of the surgical field.

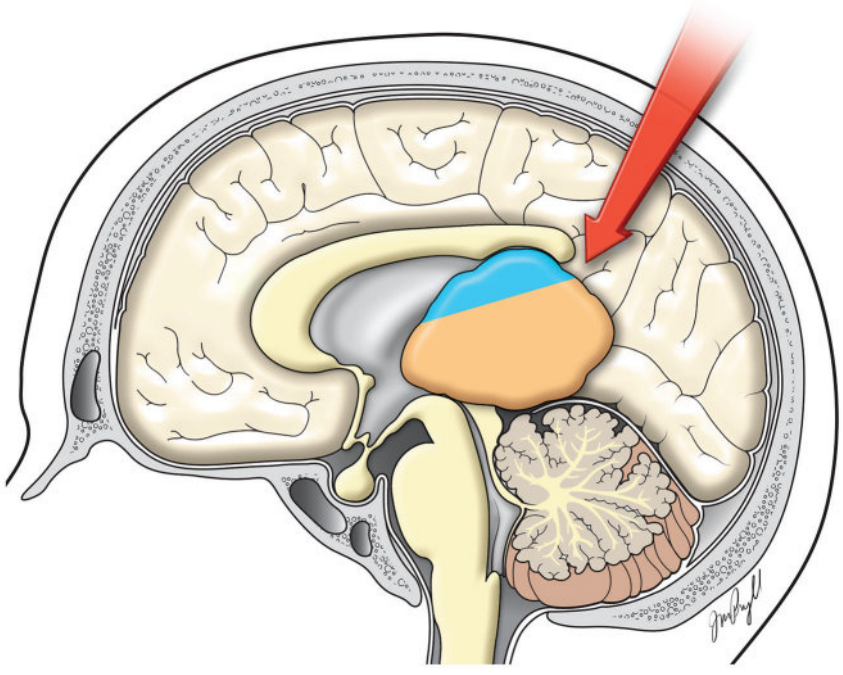


Fig. 22.9 Hidden corners. With the parietal interhemispheric retrocallosal transfalcine approach, tumor component tucked under the splenium of the corpus callosum (*colored in blue*) may be difficult to access.

■ Case Presentation 1

A 65-year-old man with a known history of a falcotentorial meningioma developed episodes of confusion and gait instability, and serial MRIs revealed growth of the tumor over a course of 5 years.

Anatomical and Therapeutic Considerations

The growth of this tumor caused compression of the dorsal midbrain and obstruction of the cerebral aqueduct with resultant hydrocephalus. There was also cerebellar edema from the tumor (**Fig. 22.10**). Due to these changes, as well as the decline in the patient's clinical status, surgery was recommended. A left-sided occipital transtentorial approach was chosen. The reason for this selection is that the base of the tumor involved the left tentorial leaf and accessing this portion of the tumor base across from the right side is not possible because the deep venous structures would block the surgeon's line of sight. A preoperative angiogram revealed that the straight sinus filled very slowly. The tumor "blush" was not impressive and no large arterial feeders were identified and therefore preoperative embolization was not performed.

Description of the Technique

At surgery, the patient was initially placed supine for inserting an external ventricular drain to control his hydrocephalus. He was then elevated into the sitting position, which was the preferred position for the senior author in his early experience. A central line was placed and a precordial Doppler was applied to detect air embolus. The patient's head was stabilized in a three-point headrest and the chin was gently flexed on the neck, avoiding extreme degrees of flexion. Neuronavigation was registered and a broad-based inverted horseshoe-shaped incision was planned to preserve the blood supply from both occipital arteries.

Neuronavigation was used to determine how wide the craniotomy had to be for adequate visualization of the contralateral border of the tumor. A close examination of the venous phase of the angiogram helped determine the superior edge of the craniotomy, which was planned up to the most posterior cortical draining vein. The inferior edge of the craniotomy was at the transverse sinus. A rectangular bone flap was used with two burr holes over the superior sagittal sinus and one burr hole just superior to the transverse sinus.

During the craniotomy, the anesthesia team alerted that the patient's airway pressures became acutely elevated. There were signs of hemodynamic instability, and the signal on the precordial Doppler was indicative of an air embolus. The field was flooded with saline, the wound was quickly closed, and the patient was lowered to a supine position. Once the patient was stabilized, the decision was made to proceed. The patient was repositioned in the prone position with the chin tucked and the head elevated above the heart. The risk of an air embolus was thus minimized, but not entirely eliminated.

The dura was opened slightly off the midline. If draining veins entering the superior sagittal sinus are encountered during this approach, they must be carefully dissected to preserve the arachnoid around it so that the occipital lobe can be mobilized laterally. This careful manipulation of the veins creates a wider working area at the midline to create a corridor down to the tumor. A lot of vigilance on the part of the surgeon is required so as not to compromise the cortical veins while working deep to them.

During the tumor resection, the occipital lobe was held away with a retractor to prevent it from sagging down with the gravity. The tentorium was divided all the way to the incisura, leaving about a centimeter edge from the straight sinus. Cutting the tentorium a sufficient distance away from the straight sinus circumvents venous lakes in the tentorium that can be troublesome. Sometimes it is difficult to identify the exact location of the straight sinus, and navigation can be very useful in this step. The usual microsurgical principles of internal debulking and capsular dissection were performed, preserving the vital neurovascular structures. In this case, the dissection was very tedious and difficult due to the adhesions between the tumor and the cerebellum. The tentorial attachment of the tumor was excised on the left side. The falx was then retracted laterally, and reaching underneath the straight sinus, tumor was delivered sequentially from the contralateral side. This access across

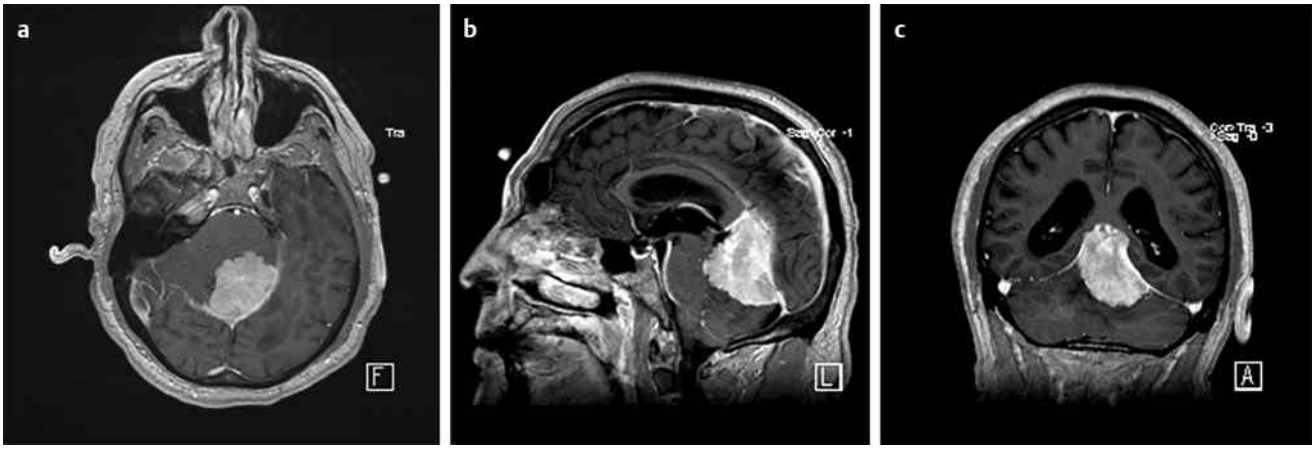


Fig. 22.10 MRI of Case 1. **(a)** Axial, **(b)** sagittal, and **(c)** coronal preoperative MRI revealing a large left falcotentorial meningioma extending both supratentorially and infratentorially and causing hydrocephalus from obstruction of the cerebral aqueduct in the dorsal aspect of the midbrain. There was also cerebellar edema from the tumor.

Fig. 22.11 Postoperative MRI of Case 1. (a) Axial, **(b)** sagittal, and **(c)** coronal images revealing gross total resection of the falcotentorial meningioma through an occipital transtentorial approach with resolution of the obstructive hydrocephalus and preservation of the deep venous structures.

to the contralateral side was limited by the space under the straight sinus and vein of Galen.

After the resection was complete, closure was performed in the usual fashion.

Aftercare

Postoperative MRI revealed gross total resection of this tumor and pathology confirmed it was a WHO grade I meningioma (**Fig. 22.11**). The patient developed a right homonymous hemianopsia which was present at 1 year. There has been no tumor recurrence in 6 years of follow-up, and he continues his work as principal of a college. He did not require permanent shunting.

■ Case Presentation 2

A 50-year-old man with a history of neurofibromatosis type II, who had multiple previous operations, presented with a progressively enlarging meningioma in the region of the pineal gland that was causing severe compression of the brainstem, cerebellum, and occipital lobes (**Fig. 22.12**).

Anatomical and Therapeutic Considerations

This meningioma was large and had a wide attachment arising at the tentorial incisura. An angiogram was done prior to surgery, which showed obstruction of multiple venous channels and multiple unnamed venous collateral channels. The deep venous system was extremely abnormal and the vein of Galen as well as the straight sinus did not fill at all. It was therefore imperative to preserve as many of the veins as possible, since there was no way of determining which channels were critical. He had a dominant right transverse sinus and the superior sagittal sinus drained into it. The left transverse sinus was small, but patent.

Given the size and vascularity of the tumor, the choice of approach was a left-sided transtentorial transsinus approach. We believed that this approach, which combined the supra- and infratentorial corridors, would give us a more direct trajectory, wider access, and more line of sight of the operative field than any of the approach options alone. The supracerebellar infratentorial approach would not have been adequate for a tumor of

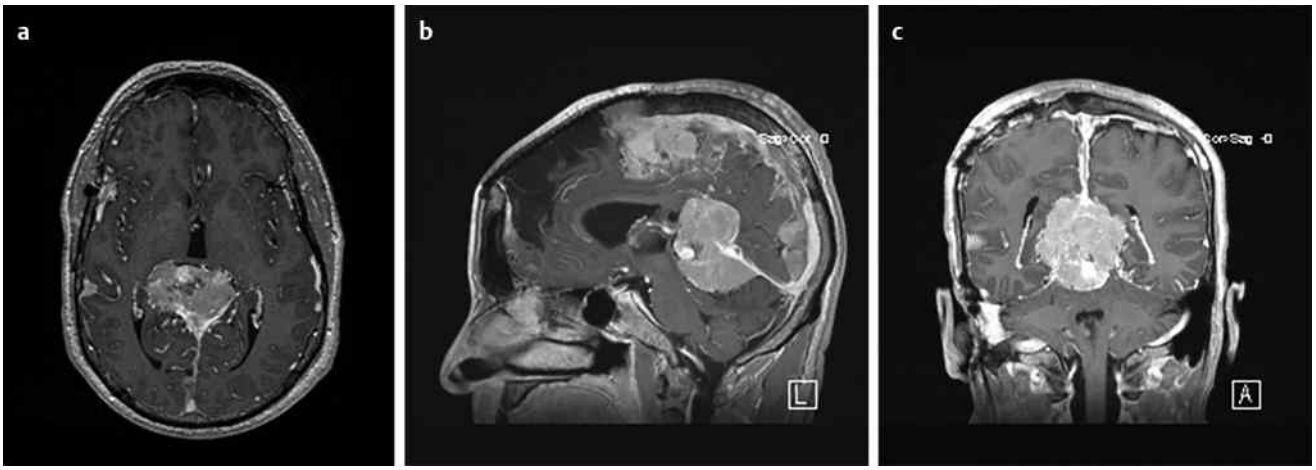


Fig. 22.12 MRI of Case 2. (a) Axial, **(b)** sagittal, and **(c)** coronal preoperative MRI revealing a large meningioma with a wide attachment arising from the tentorial incisura causing severe compression of the brainstem, cerebellum, and occipital lobes in a patient with a history of neurofibromatosis type II.

this size with a large supratentorial component. Alternatively, the biparieto-occipital approach would be inadequate for resection of the infratentorial component and would jeopardize both hemispheres.

Description of the Technique

The left-sided approach was made with the patient in lateral position and right side down. The neck was flexed so that the surgeon could look in a cephalad and caudal direction with ergonomic ease and comfort while operating. The nose of the patient was turned about 30 degrees toward the floor. The side of the approach was chosen based on the side of the nondominant sinus and for better visualization of the contralateral side where the tumor's largest component existed. A broad-based inverted horseshoe scalp flap was planned to preserve bilateral occipital artery blood supply. The medial aspect of the dependent leg was exposed and prepped for harvesting the greater saphenous vein, anterior to the medial malleolus, to repair the transverse sinus.

Navigation was used to assist with planning of the craniotomy, which straddled the transverse sinus. In the midline, it came close to the superior sagittal sinus. The superior and inferior extent of the craniotomy was planned according to the cephalad-caudad extent of the tumor to obviate the need for retraction on either the occipital lobe or cerebellum.

Dural incisions were made 1 cm above and below the transverse sinus, parallel to it. The planned site for the division of the sinus should be far enough from the torcula to protect it. The intrasinus pressure was measured by inserting a 25-gauge needle, with the tip bent to 45 degrees, into the sinus close to the sigmoid transverse junction (**Fig. 22.13a**). It was then attached to a transducer and a baseline measurement was made. A temporary aneurysm clip was placed medial to the measurement site. Since there was no change in the pressure, the sinus was deemed "safe" to divide. The temporary aneurysm clip was removed and the sinus was cut. The table was put in a slight reverse Trendelenburg position to reduce the venous bleeding. A number 3 French Fogarty balloon catheter was inserted into each end to control the bleeding (**Fig. 22.13b**), and the sinus incision was carried through the tentorium along the straight sinus all the way to the incisura. We avoided temporary clips on the cut ends of the sinus, as the clips might traumatize the thin walls.

During the tumor resection, the occipital lobe was held up with a retractor to prevent it from falling into the field (**Fig. 22.14**). The usual microsurgical principles of internal debulking and capsular dissection were performed preserving the arteries, veins, and the brainstem. Throughout the dissection, the balloon of the Fogarty catheter was deflated intermittently to allow the sinus to bleed to prevent thrombosis. Heparinized saline solution was used frequently during this time but no systemic heparin was given.

After the resection, closure began with the repair of the sinus. Direct reanastomosis was not possible because the dura shrank back. A greater saphenous vein graft was harvested from the lower leg anterior to the medial malleolus, making sure that no valves existed in the segment (**Fig. 22.13c**). 7–0 Prolene sutures were then used to suture the interposition graft in place. The Fogarty catheters were removed before the final sutures were placed and the ends were allowed to bleed. The graft was irrigated with heparinized saline to remove air bubbles and the sutures were then completed (**Fig. 22.13d**). Since the transverse sinus and the saphenous vein graft segment are without valves, patency can be checked by "milking" them in one direction and then another.

Aftercare

Postoperative MRI revealed the expected residual tumor along the posterior incisura of the tentorium (**Fig. 22.15**). It also demonstrated small ischemic insults in the superior cerebellum bilaterally, splenium, and posterior right thalamus, likely resulting from obliteration of perforating vessels that were adherent to the tumor. The patient was slow to wake up postoperatively and, when he did, had a component of dysphasia and paucity of speech akin to cerebellar mutism. This improved significantly prior to discharge and was fully resolved at his 3-week postoperative visit. He did not have any visual field deficit.

An immediate postoperative MRV was obtained with questionable patency of the saphenous vein graft, and he was

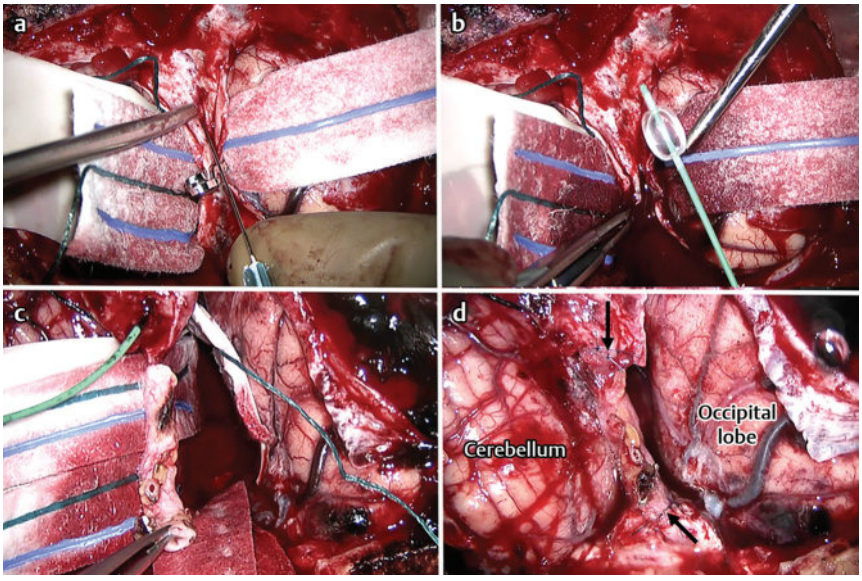


Fig. 22.13 Opening and repair of transverse sinus. **(a)** A temporary clip has been placed on the proximal left transverse sinus and the sinus pressure was measured more distally in preparation for opening the sinus. **(b)** The sinus was opened and Fogarty balloons were used to control the bleeding. **(c)** The vein graft was measured to size for use to repair the sinus. **(d)** Final view after sinus repair with the *black arrows* indicating the suture lines.

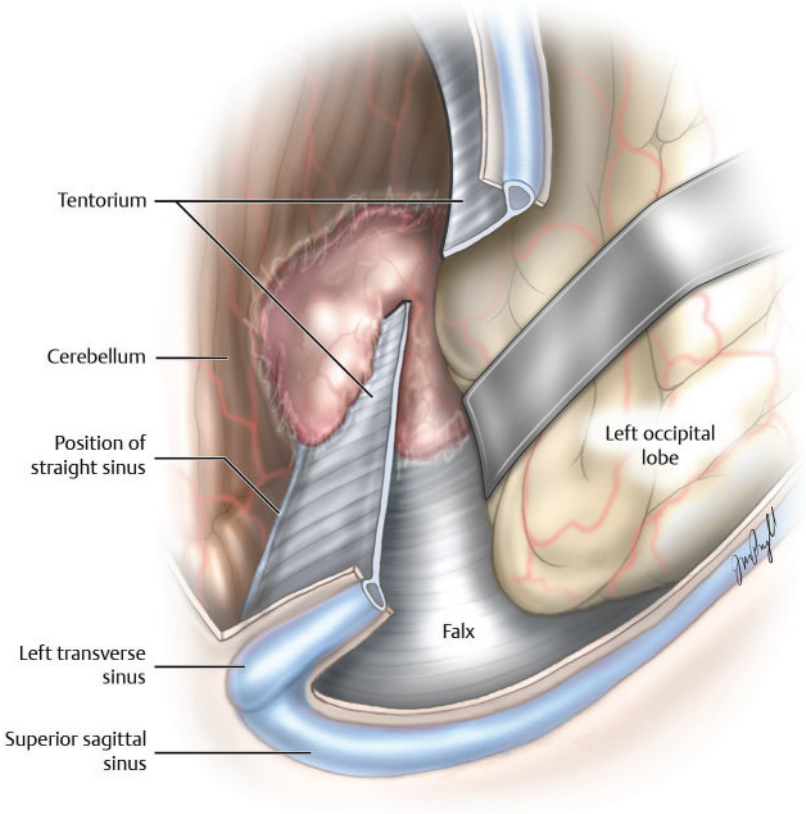


Fig. 22.14 The occipital transtentorial transsinus (combined supra- and infratentorial) approach. The addition of the posterior fossa bone opening, as well as the tentorial incision from transverse sinus to incisura, expand the surgeon's line of sight to include an inferior-to-superior trajectory, which is unavailable when the tentorium is only partially cut from above.

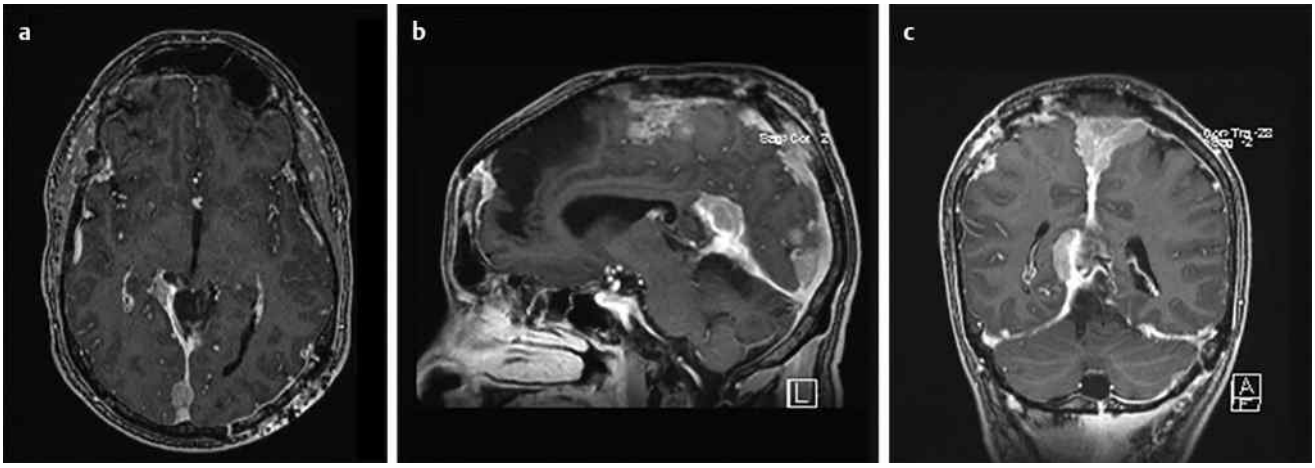


Fig. 22.15 Postoperative MRI of Case 2. (a) Axial, **(b)** sagittal, and **(c)** coronal images revealing residual enhancing tumor along the surgical cavity at the posterior incisura of the tentorium as expected, which was initially observed and later on treated with radiosurgery when it demonstrated interval growth.

therefore started on low-dose aspirin. An angiogram 1 year after surgery revealed that left transverse sinus was open, albeit with less flow than the right (**Fig. 22.16**). Histopathological analysis revealed a meningioma with atypical features but it did not meet criteria for an atypical meningioma. The tumor was carefully monitored and when it demonstrated progression, radiosurgery to the residual tumor was performed.

Commentary

We believe that the occipital transtentorial approach is an excellent option for most falcotentorial/pineal region tumors, and reserve the transtentorial transsinus approach for very large vascular tumors with significant bilateral as well as supra/infratentorial components. Each surgical option presents its

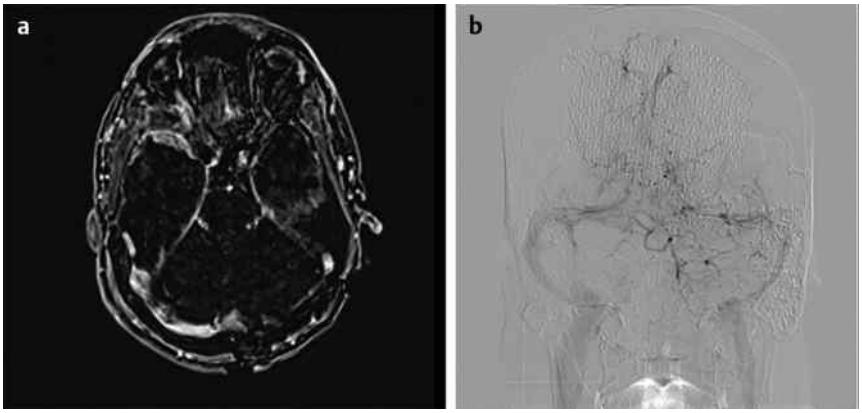


Fig. 22.16 **(a)** Immediate postoperative MRV revealing a filling defect in the region of the left transverse sinus thus putting into question the patency of the left saphenous venous graft. In order to maximize possibility of maintaining venous patency, the patient was started on antiplatelet therapy. **(b)** An angiogram 1 year postoperatively revealing that left transverse sinus was open, albeit less opacified than the right.

own set of risks and benefits, and a surgeon must choose the approach she or he believes will accomplish the surgical goal safely. Regardless of the final choice, operations for falcotentorial tumors tend to be long and tedious, and one point seldom mentioned is that the choice of *positioning* must allow the surgeon to operate comfortably for prolonged periods.

Although, by definition, operations for falcotentorial meningiomas are oncological in nature, a comprehensive understanding of the preoperative *vascular* imaging is paramount to success in these cases. As evidenced in the cases described in this section, the normal deep venous anatomy can be distorted and it is crucial for surgeons to keep their orientation in relationship to the venous structures and the collateral venous network.

Finally, one must realize the limitations of each operation and make a critical decision to stop the resection when appropriate. If the tumor is firmly attached to the critical venous structures at the depths of the surgical corridor, surgery should be halted if the primary goal has been achieved. Failure to do so can lead to catastrophic outcomes.

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23 Superior Vermis

Kyle Mueller and Walter C. Jean

Keywords: interhemispheric fissure, transtentorial approach, vermis, supracerebellar infratentorial approach, sitting position

■ Case Presentation

A 60-year-old woman presented to a community hospital with a 2-day history of gait instability and confusion. Her medical history was significant for von Hippel–Lindau (vHL) syndrome, and she had previous surgery to remove a tumor from her cerebellum. Her physical examination showed an obese woman who was slightly drowsy. Otherwise, she had no focal neurological deficits.

Magnetic resonance imaging (MRI) of the brain was obtained (**Fig. 23.1**). It demonstrated a solitary large, avidly enhancing lesion at the superior vermis. There is compression of the sylvian aqueduct with resultant hydrocephalus. She was transferred to the University Hospital's intensive care unit (ICU) for further evaluation and management.

Questions

1. Describe the genetics of vHL along with common vHL-associated clinical conditions.
2. How does the differential diagnosis of the MRI findings and patient history influence the next step in management?

■ Diagnosis and Assessment

The vHL syndrome is a multisystem neoplastic disorder that is inherited in an autosomal dominant fashion. It is caused by the inactivation of a tumor suppressor gene on chromosome 3. Commonly associated tumors include hemangioblastomas (HBLs) of various regions of the neuroaxis, renal cell carcinoma, pheochromocytoma, and endolymphatic sac tumors. About 30% of HBL are associated with vHL. These tend to present in patients about a decade earlier than sporadic HBL.

Our patient has a large superior vermian lesion that resulted in hydrocephalus, which in turn caused her to be drowsy and confused. The management of the patient should be divided into addressing her hydrocephalus and cerebellar lesion. Between the two, the hydrocephalus must be addressed first, as it, and less likely the mass lesion, can cause rapid and potentially irreversible neurological deterioration. Placement of an external ventricular drain (EVD) was performed in the ICU, and was opened at 20 cmH_2O in order to prevent upward herniation. Since we had a reliable neurological examination afterward, we decided to monitor her closely in the ICU for any acute decline, with plans for surgical resection the following day. Steroid treatment was initiated in order to reduce edema in the cerebellum and mass effect.

With regard to the cerebellar lesion, the main differential diagnosis for our patient is between HBL and metastasis. Her MRI showed a large, solid, avidly enhancing mass. Because of her history of vHL, and despite the lack of a significant cystic component, we favored the diagnosis of HBL. This suspicion was strengthened by her prior surgery, as her records indicated that her previous tumor was an HBL.

The differentiation between HBL and metastasis is important for surgical planning because of the profound vascularity of HBL. For its removal, considerations must be made similar to arteriovenous malformations, with early elimination of the arterial feeders and dissection of the lesion en bloc. For this to be possible, surgical exposure must be sufficiently wide. Piecemeal removal of an HBL, in a way metastatic lesions are most frequently handled, can lead to massive blood loss.

The diagnosis of HBL is important to consider for a cerebellar lesion in any adult, even without a history of vHL, because of the tumor vascularity. The enhancing nodule can vary in size and sometimes a cystic component is minimal. An example of such is shown in **Fig. 23.2**. This MRI belonged to an adult patient with no familial or personal history of vHL, and the cerebellar lesion had only a tiny cystic component. Preoperatively considered to be a metastasis, the tumor was approached for piecemeal resection, which resulted in a 1.5 L blood loss and

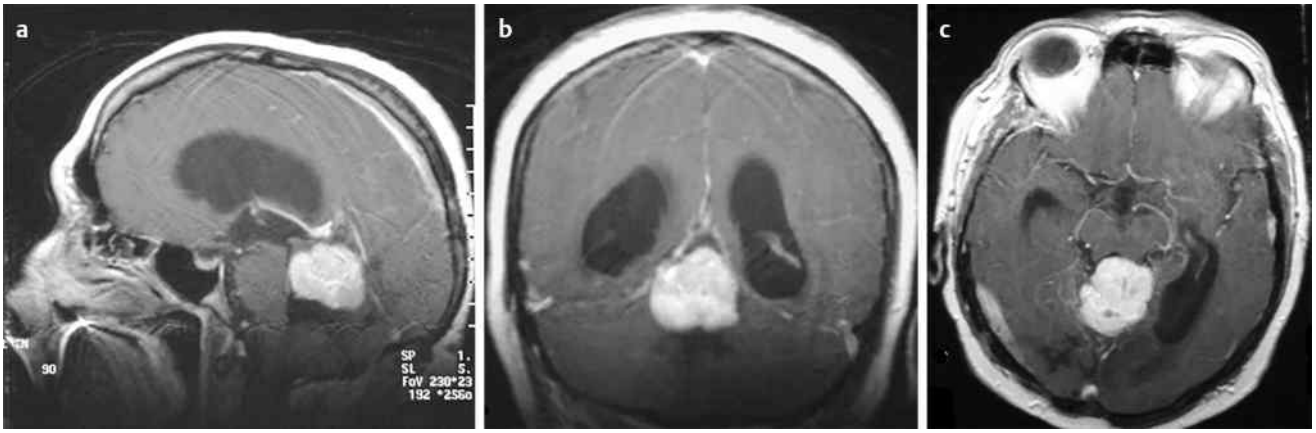


Fig. 23.1 Preoperative MRI. **(a)** Sagittal, **(b)** coronal, and **(c)** axial MRI with gadolinium demonstrating a large superior vermian enhancing lesion. There was compression of the sylvian aqueduct with resultant hydrocephalus.

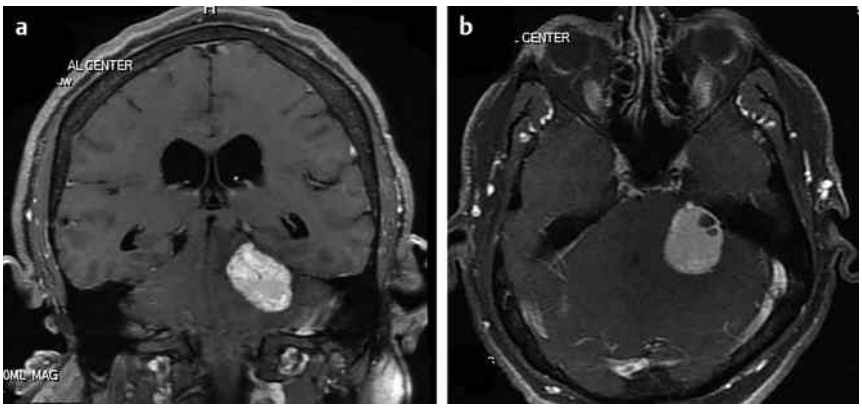


Fig. 23.2 MRI of a different adult patient. **(a)** Coronal and **(b)** axial MRI with gadolinium, showing a large enhancing lesion with a very small cystic component.

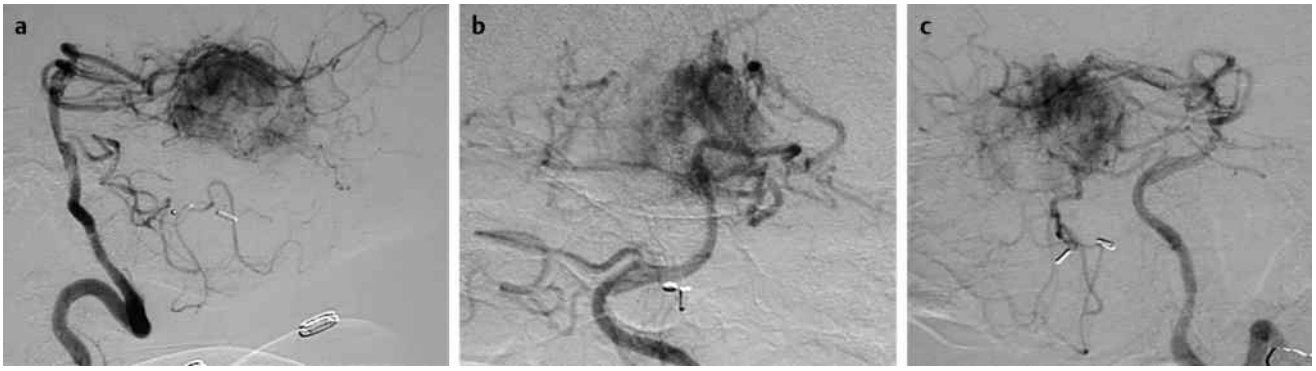


Fig. 23.3 Angiogram of the lesion. **(a, b)** Anteroposterior and **(c)** oblique projections of a selective vertebral artery injection that demonstrated a highly vascular cerebellar lesion that was primarily supplied by the superior cerebellar artery.

a much longer-than-expected operation. An angiogram before surgery would likely have changed the surgical plan.

For our patient with vHL, a cerebral angiogram was performed to add weight to the thought of HBL, assess the vascularity, and determine the source of feeding vessels. Her angiogram, as shown in **Fig. 23.3**, demonstrated a highly vascular lesion, consistent with the diagnosis of an HBL, which is primarily supplied by the superior cerebellar artery (SCA). Embolization of the arterial feeders was not feasible because of technical limitations. This left us in a position to plan surgery for a highly vascular tumor, for which we would need wide exposure to minimize blood loss.

One final thought about HBL: Many patients with vHL/HBL also have polycythemia. The two are linked because of a vHL protein product that plays a role in the oxygen-sensing pathway, resulting in upregulation of hypoxia inducible factor. Polycythemia can in turn lead to hypercoagulability, which may be important to consider before any surgical intervention. Fortunately, our patient's blood profile was normal.

■ Anatomical and Therapeutic Considerations

The cerebellum is bordered by three surfaces: petrosal, tentorial, and suboccipital, and the superior vermis is the highest point of the cerebellum, lying just under where the tentorial leaflets intersect with the falx cerebri (**Fig. 23.4**). Directly above the tentorium are the occipital lobes. The vermian surface is subdivided, from anterior to posterior, into culmen, declive, and folium. The cerebellum is supplied primarily by three cerebellar vessels: the SCA, anterior inferior cerebellar artery (AICA), and posterior inferior cerebellar artery (PICA). The vermis and tentorial surface are supplied by the SCA. The venous drainage of the area is directed anteriorly toward the vein of Galen, and eventually straight sinus.

The cerebellum is somewhat forgiving to insults such as surgical manipulation; however, one area that should be protected is the deep cerebellar nuclei. The dentate nucleus, the most lateral of the group of four nuclei (fastigial, emboliform, globose, dentate), is supplied by the SCA, and damage to this nucleus has been implicated in cerebellar mutism. Midline suboccipital and infratentorial, supracerebellar approaches have higher risks of this complication. The trochlear nerve is the only cranial nerve that exits the brainstem dorsally. Given the location of our patient's lesion, special care must be taken at the anterior edge of the tumor to limit any postoperative visual complications from injury to the trochlear nerve.

The large enhancing nodule in our patient was located in the superior vermis, and as previously mentioned, the surgical approach warrants special consideration between of the unique features of HBLs. The trajectory should be straight and the corridor sufficiently wide for the resection to be with minimal violation of the tumor capsule. Ideally, the approach should also allow early access to the arterial feeders from SCA, so that these can be eliminated promptly to minimize blood loss.

Options for Approach

The midline, suboccipital corridor is the "work-horse" approach used most frequently to remove midline cerebellar mass lesions.

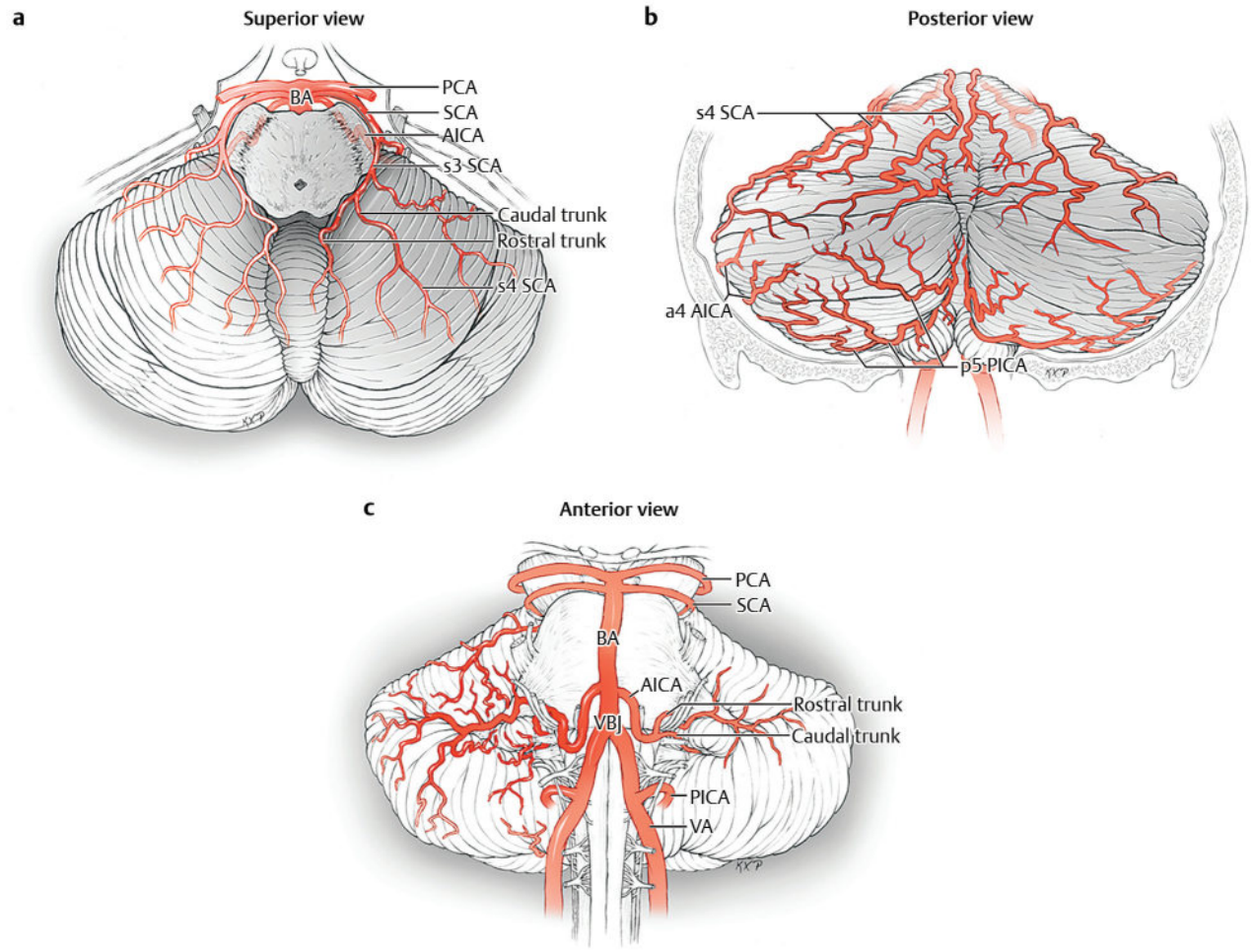


Fig. 23.4 Cerebellar surfaces and anatomy of the cerebellar arteries. (a) The tentorial surface is supplied by cortical branches of the superior cerebellar artery (SCA) (superior view). **(b)** The suboccipital surface is supplied by the posterior inferior cerebellar artery inferiorly (PICA) (posterior view). **(c)** The petrosal surface of cerebellum is supplied by the cortical branches of the anterior inferior cerebellar artery (AICA) (anterior view). BA, basilar artery; PCA, posterior cerebral artery; s3, cerebellomesencephalic segment; a4/s4/p5, cortical segment; VA, vertebral artery; VBJ, vertebrobasilar junction. (Reproduced from Lawton M. Seven AVMs. Tenets and Techniques for Resection. 1st ed. Thieme; 2014.)

For deep-seated tumors, the transvermian route may be "added on" to reach the target, but for our patient, the modification would be an infratentorial, supracerebellar "climb" toward the target at the superior vermis. Flexing the neck and using a sitting position can mitigate the steepness of this climb, but with our patient's large body habitus, this was technically difficult and potentially hazardous.

There is currently no agreement in the literature on a method to measure and describe the "angle of the tentorium," nor is there a threshold to consider what angle is "steep." An angle is defined by two lines, and the main discrepancy is which line to use against the tentorium to define the angle. Various investigators have used different reference lines to measure the tentorial angle, but we believe that a reliable method should utilize posterior fossa landmarks for this. As such, our preferred way to measure the tentorial angle is to use the line perpendicular to the floor of the fourth ventricle as the reference line (**Fig. 23.5**). In a study with 240 subjects, we determined that the tentorial angle ranges from 43 to 86 degrees in the general population, with a mean of 60 degrees. Any angle above this, the mean would be considered "steep." This patient's tentorial angle is 67 degrees, and thus her anatomy was therefore considered challenging for an inferior-to-superior trajectory.

Approach of Choice

An alternative way to approach our patient's HBL is to use a superior-to-inferior trajectory, via the occipital interhemispheric transtentorial approach (OITA). The weakness of the infratentorial supracerebellar approach coincides with the strength of the OITA, as with the transtentorial route, a steep tentorial angle actually brings the operative target closer to the surgeon. Furthermore, the trajectory is more direct because the OITA provides a more perpendicular angle of attack. As soon as the tentorium is incised, the wide field also allows access to the anterior aspect of the tumor where the majority of feeding vessels originate. These same vessels would have been hidden from the surgeon behind the tumor in an infratentorial approach. Finally, the OITA can be performed with this large patient in a comfortable and safe lateral position, eliminating the air embolism concerns of the sitting position.

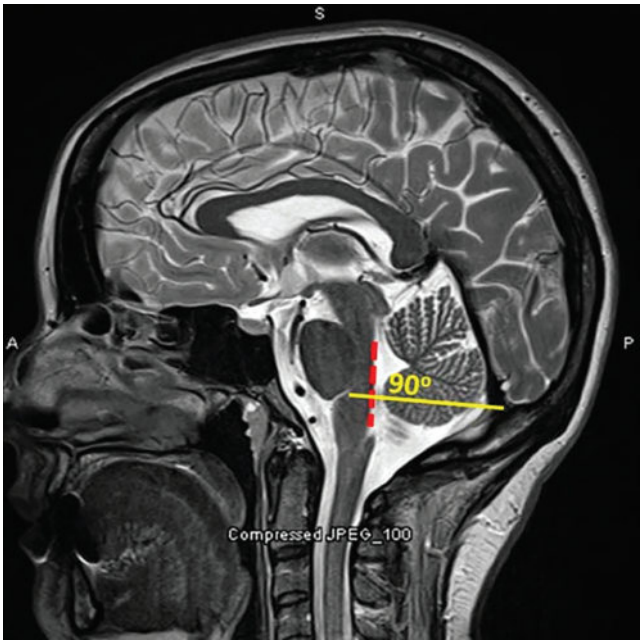


Fig. 23.5 Measuring the tentorial angle. The floor of the fourth ventricle is marked by the line *in red*, extending inferiorly from just under the inferior colliculus. The new reference line, *in yellow*, is drawn perpendicular to this red line, extending through the torcula. The tentorial angle is measured against this yellow line.

The main risk of the OITA for this patient is to the healthy occipital lobe on the side of the approach. Despite placing the ipsilateral down in the lateral position and using gravity retraction to allow the occipital lobe to "fall away" from the surgical field, there remains a risk to the patient's vision. Furthermore, it is important to note that the OITA is a unilateral approach. For any tumor that extends significantly laterally on the contralateral side, the OITA may be insufficient to reach the contralateral pole.

Despite these potential drawbacks, the OITA was the best option for our patient. Because of her body habitus, the steep tentorium and the difficulty to reach the anteriorly located arterial feeders, the suboccipital corridor was suboptimal. In addition, her previous cerebellar surgery was through the suboccipital route and scar tissue would make the dissection more difficult. For these reasons, the decision was to use the OITA to approach her HBL from superior-to-inferior, with the goal of total resection.

Questions

1. Before one cuts the tentorium, what structure must be carefully identified?
2. Is there a way to slightly improve the visualization of the contralateral side during the OITA?
3. Can you think of any technical nuances to reduce the risk for procedure-related visual complications when utilizing the OITA?

■ Description of the Technique

The patient was positioned in a lateral position with the ipsilateral (left) side down. The decision to use the left side for the approach was based on her tumor being slightly larger on the left side. After padding her axilla to prevent compression neuropathy, her head was secured with a three-point fixation using a Mayfield head holder. The neck was placed in a completely neutral position without flexion or rotation to either side. It was critical to secure her body with additional straps and tape, not only because of her weight, but also because it was predictably necessary to tilt the table during the long operation (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: interhemispheric

Craniotomy: occipital

Modifiers: incision of the tentorium

The transverse sinus was approximated using external landmarks and neuronavigation. The planned incision started in the midline about 2 cm caudal to the torcula and carried superiorly, curving over the left parietal boss in a "hockey-stick" fashion. The skin, galea, and pericranium were reflected inferolaterally. Stereotactic neuronavigation was used to identify the transverse and superior sagittal sinus (SSS). Two burr holes were made in the midline on the SSS, with two additional ones more laterally. A rectangular bone flap (approximately 5 × 3 cm) was elevated with utmost care not to violate the SSS. The dura was opened with a "U" shape based *laterally*, away from the SSS (see **Operative Setup**).

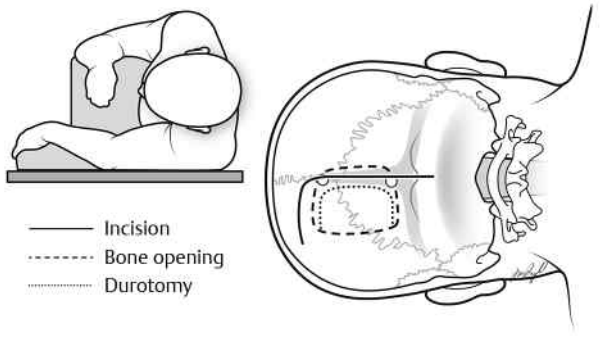
Operative Setup

Position: lateral with ipsilateral side down

Incision: "hockey-stick" starting midline inferior to the inion and curved over the parietal boss

Bone Opening: occipital craniotomy with midline burr holes on the SSS

Durotomy: "U" shaped based laterally with an incision close to and parallel with the SSS



As gravity retraction mobilized the ipsilateral occipital lobe laterally, dissection of the interhemispheric fissure proceeded with caution so as not to damage cortical veins. As predicted, no bridging veins from the cortex to SSS were encountered. The falx was clearly identified and a retractor blade was placed here to gain slightly more exposure of the right side. This retraction was against gravity, but fortunately, the falx protected the right occipital lobe reliably well. The falx was then

followed inferiorly to the tentorium. At this point, it was critical to identify the straight sinus before any additional incisions are made (**Fig. 23.6b**). Neuronavigation (and even fluorescein and indocyanine green [ICG]) can sometimes help, but we were able to identify the straight sinus visually without difficulty. The tentorial incision was made just lateral to the straight sinus from approximately midpoint of the straight sinus all the way to the tentorial incisura (**Fig. 23.6c**). Bipolar cautery and small hemaclips were used to stop the venous bleeding found along the way.

Once the tentorium was cut, exposure was gained to the tentorial surface of the left cerebellum (**Fig. 23.6d**). The tumor appeared as a reddish-purple mass just under the tentorium. Care was taken not to violate the capsule and the tumor was dissected away from the cerebellum in a circumferential manner (**Fig. 23.7**). Feeding arteries were found anteriorly as predicted, and were coagulated and detached. Fortunately, we had no difficulty reaching the contralateral surface of the tumor, and we did not encounter any major bleeding while moving around the edges of the mass. Thus, the tumor was removed en bloc (**Fig. 23.8**)

The cut edges of the tentorium were left as closure began. Since the cut started only at the midpoint of the straight sinus, we were not concerned about any occipital lobe "sag" into the posterior fossa. The dura was closed in a watertight fashion with 4–0 Nurolon suture and the bone flap was replaced and secured to the cranium. The scalp was closed in a standard fashion. The patient was extubated and transferred to the ICU with the EVD under continual drainage.

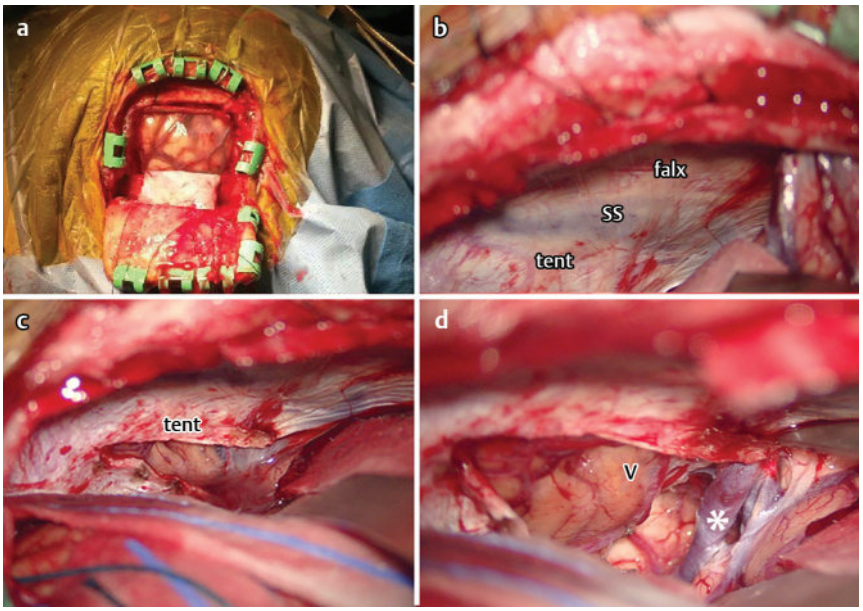


Fig. 23.6 Intraoperative illustration: occipital transtentorial approach to the cerebellar vermis. A right-sided approach is shown in these images of a different patient. **(a)** This patient is in a right-sided down lateral position and a right-sided craniotomy exposed the right occipital lobe. Note the dura is flapped away from the midline and superior sagittal sinus to avoid a "free edge" against which the occipital lobe can fall. **(b)** The falx blends into the tentorium (tent), and the straight sinus (SS) is the only demarcation. **(c)** The tentorium is cut lateral to the SS from midway to the incisura (tent: cut edge). **(d)** This cut exposed the cerebellar vermis (V). The vein of Galen (*) is seen joining the straight sinus.

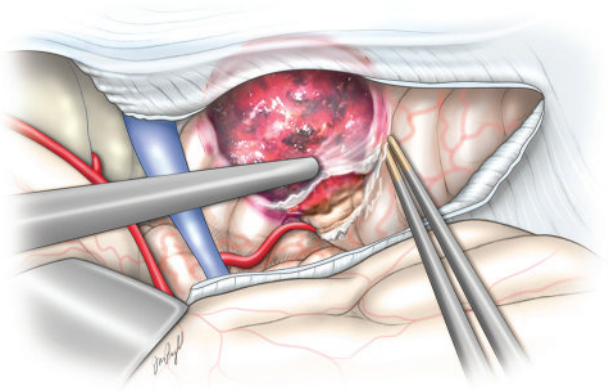


Fig. 23.7 Illustration of the occipital interhemispheric transtentorial approach. This depicts the resection of the vermian hemangioblastoma in the patient described in this chapter. The patient had a steep tentorium, and the transtentorial approach utilizing the patient's left side was performed for complete removal of the lesion.

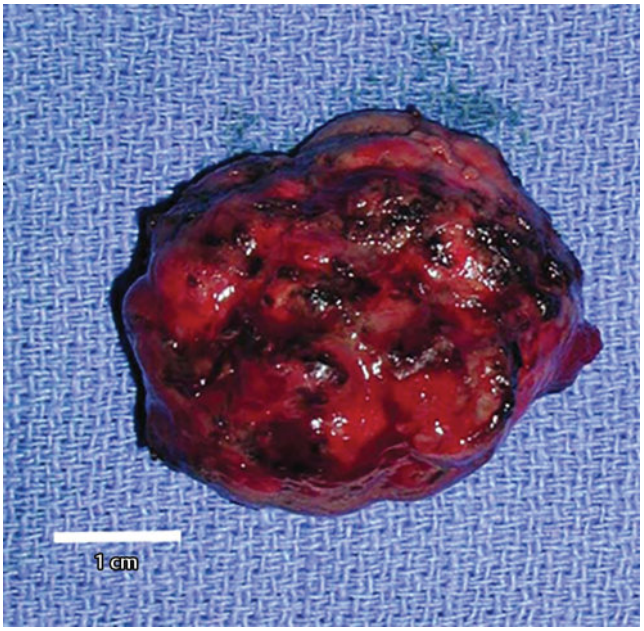


Fig. 23.8 The gross pathological specimen of the resected hemangioblastoma.

Surgical Pearls

1. The two midline burr holes should be carefully made over the SSS, so as not to injure the sinus. Once made, these burr holes allow careful dissection of the SSS away from the inner table, and facilitate the craniotomy cut with the drill to be placed precisely above the contralateral (right) edge of the SSS. The placement of this cut then allows the falx to be mobilized slightly to the right once the operation proceeds intradurally.
2. The dura was opened with a "U" shape based laterally, away from the SSS. This is against "conventional wisdom," as most surgeons would base this cut along the SSS. We have found that in this more traditional manner, gravity would pull the occipital lobe against the "cut edge" of the dura laterally and cause significant swelling in the lobe. As bridging veins between the occipital lobe and SSS scarcely exist posterior to the vein of Trolard, cutting the dura right next to the SSS prevents this occipital lobe "strangulation." Furthermore, with the dural flap based laterally, the flap itself can sometimes be used to protect the occipital lobe from the abrasion of tools moving in-and-out of the field.

■ Aftercare

Our patient tolerated the procedure without any complications. She was extubated and returned to the ICU. Her postoperative MRI demonstrated a total resection of the tumor (**Fig. 23.9**). She continued to be monitored in the ICU and her steroids were weaned off. Deep venous thromboembolism prophylaxis was initiated on postoperative day 1. Histopathological analysis confirmed the lesion to be a HBL.

The EVD was used during the postoperative period to evacuate any surgical debris and blood products circulating with the cerebrospinal fluid (CSF). After much of this was cleared, the EVD was weaned by progressively raising it to lower the daily output. However, this patient ultimately required placement of a ventriculoperitoneal shunt because of failure to wean the EVD. She experienced a left-sided homonymous hemianopsia which improved gradually. She was discharged to rehabilitation. Thereafter, she was monitored with yearly MRI scans for recurrence. At her last follow-up, more than 5 years after the surgery, no recurrence has been detected. Her left-sided vision had improved to a point where it was not noticeable to her or to testing.

■ Possible Complications and Associated Management

A variety of complications can occur that are specific to posterior fossa approaches. CSF leakage, hemorrhage, hydrocephalus, cerebellar mutism, and visual disturbances are all possibilities that the surgeon must consider and counsel the patient on.

As with any posterior fossa operation, CSF leakage is a concern. Meticulous dural closure, the generous use of dural substitute and sealant, and maintaining the head of bed above 30 degrees postoperatively can all reduce the incidence of leakage.

Cerebellar mutism is a feared complication that can affect pediatric and adult patients alike. Avoiding damage to the cerebellar vermis and dentate nucleus can minimize the chances of this complication. Preoperative counseling and postoperative reassurance are important to the patients and their families, as most often, mutism is transient.

Although our patient did not have any permanent visual disturbances, injury to the trochlear nerve as well as the occipital lobe can occur. With the OITA, proper positioning to allow gravity retraction is critical. During the surgery, attention is inevitably focused on the target deep in the surgical field. It is nevertheless important to look at the occipital lobe time-and-again to make sure that it is not in harm's way. Drainage of CSF via a lumbar drain is another option to consider in order to increase brain relaxation. Injury to the trochlear nerve can occur with approaches around this area. Given the small size of the nerve and its location deep to the tumor, heightened awareness to seek out the nerve is often the only way to preserve it during tumor dissection.

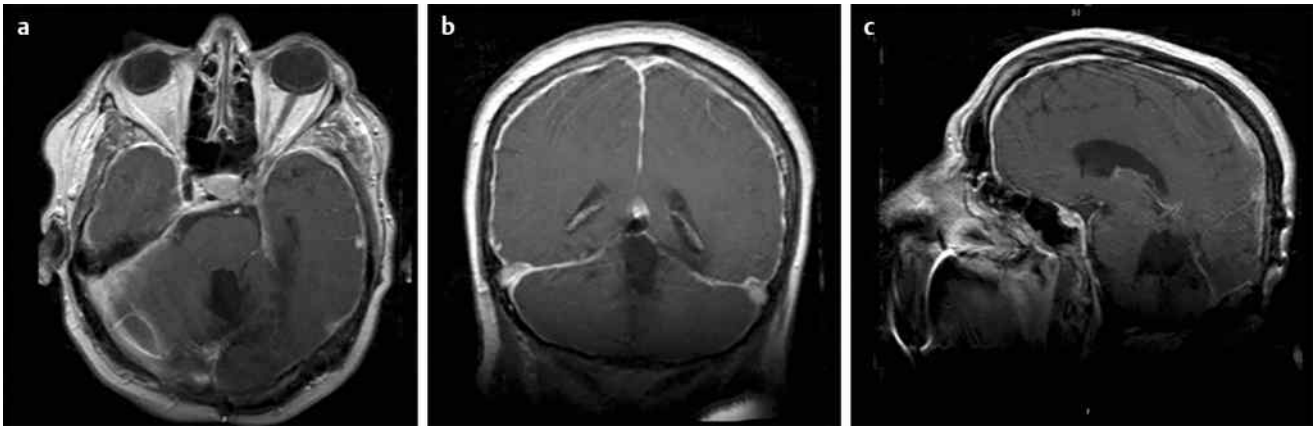


Fig. 23.9 Postoperative MRI. **(a)** Axial, **(b)** coronal, and **(c)** sagittal postoperative MRI with gadolinium demonstrating a total resection of the prior vermian hemangioblastoma.

Perspective

Roberto C. Heros

■ Occipital Interhemispheric Transtentorial Approach

The OITA has been beautifully described in the preceding section. The authors were faced with a significant surgical challenge, presented by a large HBL of the anterior vermis of the cerebellum in a patient with vHL syndrome. The rich vascularity of these tumors is of course well-known to neurosurgeons, and this accounts for the main surgical challenge with this particular case. I will begin by saying that I agree 100% with the surgical approach that the authors selected for this case. They described meticulously the factors that led them to choose the OITA approach for this tumor. The patient had a relatively steep tentorial angle and had a large body habitus which, as the authors pointed out, makes the alternative approach, the infratentorial supracerebellar approach, about which I will comment later, more difficult. In addition, and most importantly, with the OITA approach they had a good view of the anterior aspect of the tumor where the feeding superior cerebellar branches came to the tumor and could be controlled before beginning to dissect the tumor. This is critical because these tumors should be removed in one piece after appropriate devascularization if at all possible.

I will briefly add some comments to the authors' detailed description of the operation. I well remember the days when we used to do this approach with the patient in a semi-sitting position and it was almost to be expected to have a visual field defect from the need to retract the occipital lobe in this position. This led many neurosurgeons to adopt a lateral position with the ipsilateral side down which practically eliminates the need for any occipital lobe retraction since the ipsilateral occipital lobe "falls away" by gravity. This can be helped by CSF drainage which I routinely use for this position through a lumbar drain. This was of course unnecessary in the authors' case, since the patient already had an EVD, which could be used for this purpose. The authors' show a "hockey-stick" incision but the alternative, which I prefer, is a long straight parasagittal incision which in my opinion heals better and is sufficient since the bony exposure to the ipsilateral side does not need to be more than 3 to 4 cm in the transverse diameter. I found it very interesting that the authors opened the dura with a window based laterally as opposed to the usual window based medially on the SSS, which is traditionally used for this approach. The reason for the use of a laterally based flap is to prevent damage to the occipital lobe as it falls against the sharp cut edge of the dura when a medially based dural window is used. Avoiding such damage to the brain, by its falling against the cut edge of the dura, is extremely important and is a point I have emphasized for years to our residents. The way I avoid this when using a medially based dural window is by making the dural flap relatively shallow such that as the brain falls away it falls under the dura rather than against its cut edge. It is not necessary and it is in fact contraindicated, to have a large dura window in the transverse diameter since all that is necessary is a couple of centimeters of exposure between the falx and the brain.

Generally, in parasagittal approaches I use a relatively wide dura window in the sagittal diameter so that I can select a point of entry between draining veins and avoid the sacrifice or damage to any of these veins. However, as pointed out by the authors, in the occipital parasagittal approach there are practically no veins of any importance. Those veins become more important as one moves upward toward the postcentral region which is unnecessary for this approach. Incidentally, I note that the authors use two burr holes right over the SSS for the medial aspect of the bone flap. Placing the burr holes right over the sinus is a well-accepted and time-tested technique; however, since I always fear that the worst could happen, I prefer to avoid the risk of injuring the sinus with the burr holes and place my burr holes for the medial aspect of the flap on the opposite side of the SSS. This also makes it easier, once the dura is open, to retract slightly the SSS and the falx to enhance the exposure. Once the dura is open and relaxation of the brain is obtained by CSF drainage and mannitol, the next step is to identify the straight sinus. In the authors' case they had no difficulty with this but I must admit that I have had a few cases where this has not been straightforward since frequently the angle between the falx and tentorium is slightly curved and not the usual right angle that is frequently illustrated in surgical atlases. The authors pointed out that neuronavigation and the use of fluorescein ICG can be useful to precisely identify the position of the straight sinus which of course is critical. I will add that a Doppler probe can also be useful for this purpose.

The authors started the tentorial incision, which is best done right next to the straight sinus, at the midpoint of the tentorium. I prefer to start the incision further back, just in front of the transverse sinus, to allow a wider exposure once the tentorium is completely divided. I always teach that a mistake in opening the tentorium, whether in this location or with a subtemporal approach, is to start layer by layer because when the tentorium is divided in this fashion, one may enter one of the tentorial venous channels which can make bleeding difficult to control if one has "unroofed" such venous channel without having gone through the deeper layer of the tentorium. That is why I make my cut completely "through-and-through" and then advance toward the tentorial incisura by coagulating both leaves of the tentorium simultaneously and using hemoclips when necessary. I make a point of identifying the fourth nerve before finishing my cut through the incisura to make sure that the nerve is not cut with this final incision. The next step is control of the arterial feeders as described by the authors and after that the en bloc resection of the tumor should proceed satisfactorily as it did in the authors' hands.

■ Case Presentation

An alternative approach to the anterior vermis of the cerebellum is the infratentorial supracerebellar approach. Of course, we use that approach most commonly for tumors of the pineal region but it can also be used for cerebellar lesions involving the superior (tentorial) surface of the cerebellum and the

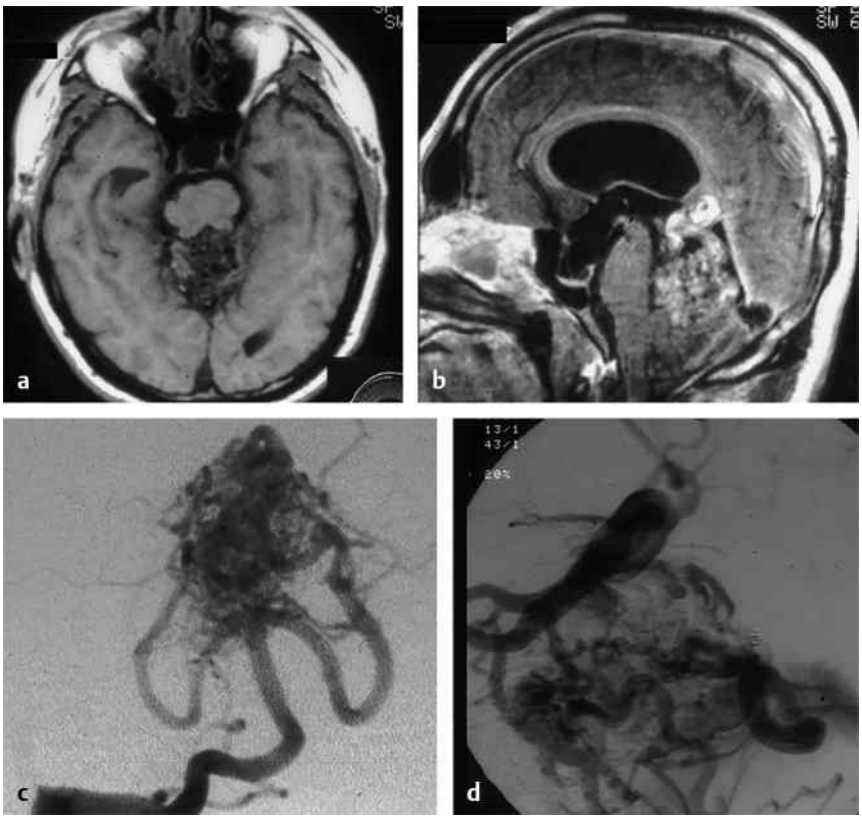


Fig. 23.10 Preoperative images. (a) Axial and **(b)** sagittal MRI of a 44-year-old man with ataxia and cognitive difficulties. **(c,d)** Digital subtraction angiogram. **(c)** Anteroposterior and **(d)** lateral images of the same patient's arteriovenous malformation.

cerebello-mesencephalic cleft. A cerebellar arteriovenous malformation (AVM) involving the anterior vermis is shown in **Fig. 23.10**. This was a patient in his 40s, who had been deteriorating neurologically mostly because of hydrocephalus. The lesion was mostly fed by the SCAs from both sides. This lesion was wider in diameter than the HBL illustrated by the authors of this chapter. Although the ipsilateral half of the lesion could have been exposed very nicely with the OITA approach, I felt that it would be difficult to reach the contralateral extent of the lesion by that approach (**Fig. 23.10a, b**). In addition, the tentorial angle in this particular case was not unusually steep and the patient had a normal body habitus which made the sitting position satisfactory. I should comment that I operated on this patient over 20 years ago and perhaps at that time I was not as comfortable with preoperative embolization and I went ahead with the surgical excision of this AVM without embolization. If I were to operate on that lesion today, I certainly would proceed with preoperative embolization to facilitate the surgery and make it less risky.

■ Description of the Technique

I will make some points, all of which are relatively well-known, about the sitting position. Some of the precautionary measures that I will mention I learned "the hard way" by operating on a patient with a HBL of the brainstem, when I first arrived to Miami. I was used to frequently using the sitting position for pineal tumors in Boston and depended on the anesthesiologist to take all the precautions that are necessary for this position. Unfortunately, the patient that I am referring to was an older man who, unbeknownst to me, had asymptomatic but severe cervical spondylosis. We proceeded to sit the patient up without monitoring sensory, motor and brainstem potentials, a precaution that I have taken ever since in every patient in the sitting position and which probably would have prevented the ensuing catastrophe. As we sat the patient up I did not tell the anesthesiologist to move the transducer recording the arterial pressure to the level of the head which resulted in blood pressure readings that were falsely higher than they would have been at the level of the brain which is where it was needed. I also did not ask the anesthesiologist to "fill the tank" with a couple of units of Albumisol before we started sitting up the patient. Now I insist on a central venous pressure (CVP) of at least 6 to 8 before we begin to sit the patient up. I also failed to insist to the anesthesiologist that the blood pressure be kept at no less than the preoperative level at all times and that I should be informed if for any reason the pressure dropped below this level. We had a significant period of hypotension during the operation with undoubtedly contributed to the problem. I did take the precaution of having a space of two fingers between the chin and the chest but that may have been too much flexion on a patient with cervical spondylosis. This patient awoke from surgery quadriplegic from a major infarction of the cervical cord which we attributed to relative hypotension in the setting of neck flexion with underlying severe spondylosis. Ever since I take responsibility on being intimately involved in the process of sitting up the patient, doing so gradually while I personally monitor the arterial pressure and confirm that the arterial pressure transducer is gradually being raised to the level of the head. I also measure evoked potentials in all of these patients and make sure that they do not change as we sit the patient up. As mentioned I insist on a CVP of at least 6 to 8 cm and keep the separate transducer for the CVP at the level of the heart.

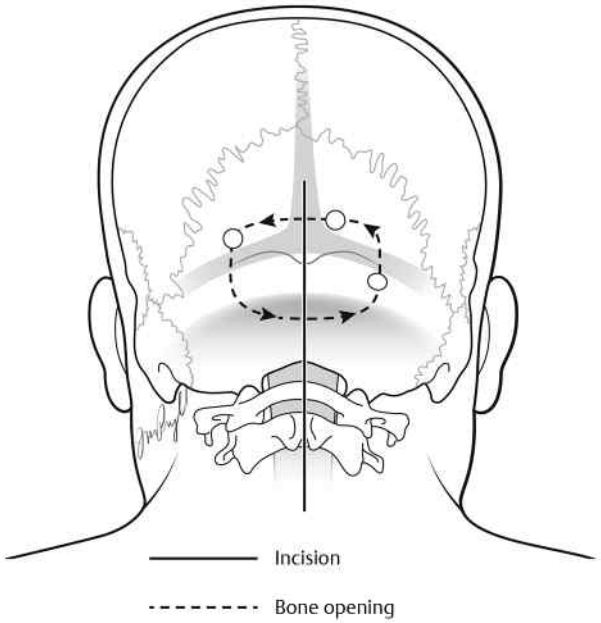


Fig. 23.11 Illustration of the placement of the burr holes and direction of bone cuts made.

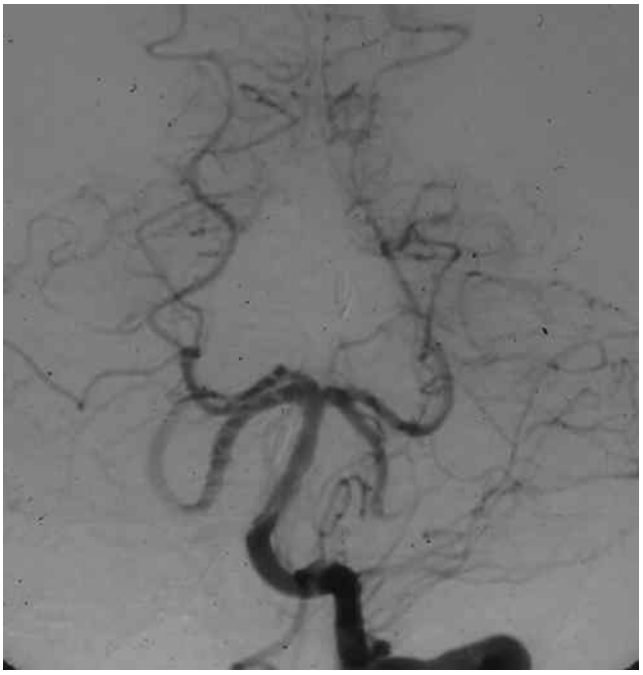


Fig. 23.12 Postoperative image. Digital subtraction angiogram after surgery showed a complete removal of the patient's arteriovenous malformation.

I repeat many times to the anesthesia team that the pressure must be kept up and that I must be notified any time that the pressure falls below the preoperative level. Needless to say, the usual precautions to prevent air embolism (a central line with position confirmed by a chest X-ray and a functioning precordial Doppler) are essential.

The bone flap that I like to use for the infratentorial supracerebellar approach is a combined occipital and suboccipital flap that extends from just above the transverse sinuses and torcula and does not reach the foramen magnum in the suboccipital region. It is important not to cut the bone flap in the suboccipital region too low to prevent the cerebellum from "falling out" of the wound over the bone flap. I am old-fashioned and insist on a single burr hole just before each sinus is crossed. After the burr hole is made, the adjacent sinus is separated from the inner table of the skull under direct vision before it is crossed with the craniotome (**Fig. 23.11**). I know that there are sleeker and faster ways to do this, frequently through just one small hole sufficiently wide to place the foot of the craniotome and then depending on the foot of the craniotome to separate the dura from the bone. However, I prefer to be conservative and avoid the one complication that could kill the patient, which is a major disruption of one of the great venous sinuses, particularly the SSS at this level. After careful separation of the dura the bone flap is removed and then the dura is opened on a window based on the transverse sinuses and the torcula. By having the craniotomy above the transverse sinus and the torcula, the dura can then be tented up forcefully to small drill holes in the edge of the bone to afford a better exposure over the cerebellum. After opening the dura, I go down to the cisterna magna to draw enough CSF in order to obtain maximal cerebellar relaxation. Almost always, the small bridging veins that hold the cerebellum up against the tentorium in the midline can be taken without serious consequence although I remember two to three cases where there was such a large prominent vein that I did not dare to sacrifice it and had to work from one side or the other of the vein. For this particular AVM, the initial step was to find and divide the large feeders from the SCA on each side (**Fig. 23.10c, d**). Since there was essentially no supply from the AICAs and only a few easily controllable small branches from the PICAs that came from below at the midline, the surgery was straightforward after arterial devascularization and the postoperative angiogram confirmed complete removal (**Fig. 23.12**).

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24 Pineal

Daniel R. Felbaum and Walter C. Jean

Keywords: tectum, quadrigeminal cistern, tentorium, endoscope

■ Case Presentation

A 22-year-old previously healthy man presented to a community hospital with a 1-month history of headache, and sudden onset nausea and vomiting. The headaches were gradual in onset, and the family had noticed some vague cognitive changes, including forgetfulness, during the last month. On neurological examination, he was found to have papilledema. A subsequent noncontrast computed tomography (CT) scan showed hydrocephalus and suggestions of a pineal region mass. The patient was immediately transferred to the University Hospital. On arrival to the University Hospital's intensive care unit (ICU), an external ventricular drain (EVD) was placed emergently to control his intracranial pressure and then a brain magnetic resonance imaging (MRI) was obtained (**Fig. 24.1**).

Questions

1. What is the differential diagnosis of the MRI finding?
2. How does the differential diagnosis influence the next step in management?
3. In addition to controlling his hydrocephalus, how can the EVD be utilized to help the patient?

■ Diagnosis and Assessment

The MRI showed an enhancing pineal region mass, and the cerebrospinal fluid (CSF) from the EVD was sent for alpha-fetoprotein and beta-human chorionic gonadotropin. The differential diagnosis for the tumor includes germ cell tumors and a wide variety of other tumor types, including pineal parenchymal tumors (PPTs), glial cell tumors, meningioma, metastasis, and miscellaneous other tumor types. Both tumor markers were negative in this patient. Had the tumor marker been positive, it would have confirmed a malignant germ cell tumor and further surgical considerations can be abandoned. These tumors are better treated with chemotherapy and radiation. The absence of tumor markers, as with this patient, does not rule out a germ cell tumor and thus it mandates a biopsy to guide further management.

The decision was made to perform a simultaneous endoscopic third ventriculostomy (ETV) and biopsy. This would yield clues of the histopathological identity of the tumor and address the hydrocephalus at the same time. In situations such as this, the choice to use a single burr hole for both purposes, versus using two separate entry sites, should be based on ventricular size, the width of the foramen of Monro, and the tumor location and size. The ideal trajectory for biopsy in the pineal region is through a frontal entry point, anterior to the standard burr hole used for ventriculostomies (Kocher's point), through the center of the foramen of Monro (**Fig. 24.2**). The ideal trajectory for performing an ETV is along a line drawn back to the surface from the tuber cinereum, up through the foramen of Monro to the surface of the skull. In this patient, the ventricles were large and the tumor was sufficiently anterior, and in general when this is the case, a single entry hole anterior to Kocher's point can achieve both goals as long as one is careful not to stretch the fornix or damage the thalamus.

A rigid endoscope was inserted through a single-entry burr hole, which was placed 1.5 cm anterior to Kocher's point and the ETV and biopsy were successfully accomplished. The histopathological diagnosis from the biopsy sample was "PPT," but further subtyping was not possible given the small biopsy sample size. A CT venogram was subsequently obtained to assess the position of the deep venous system and its relationship with the tumor mass, in preparation for further surgical intervention.

■ Anatomical and Therapeutic Considerations

Since the biopsy yielded the diagnosis of PPT with unspecified grade, the decision at this point is whether to resect the

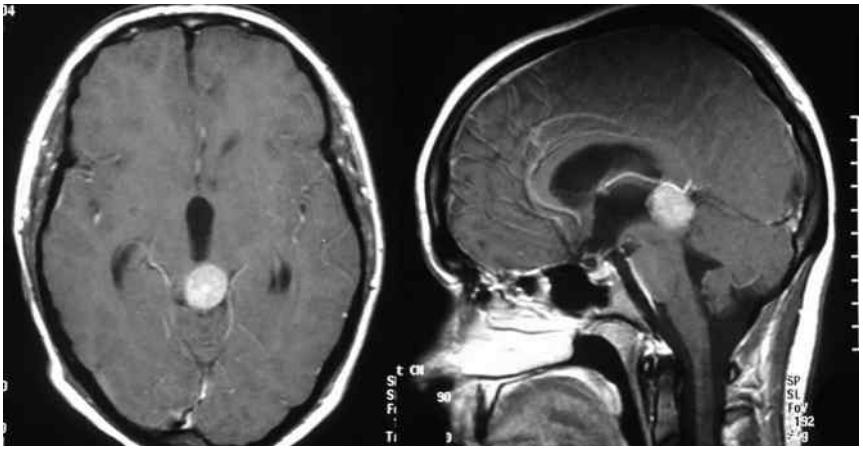


Fig. 24.1 Preoperative images. Axial and sagittal MRI images with gadolinium showed a brightly enhancing mass in the pineal region.

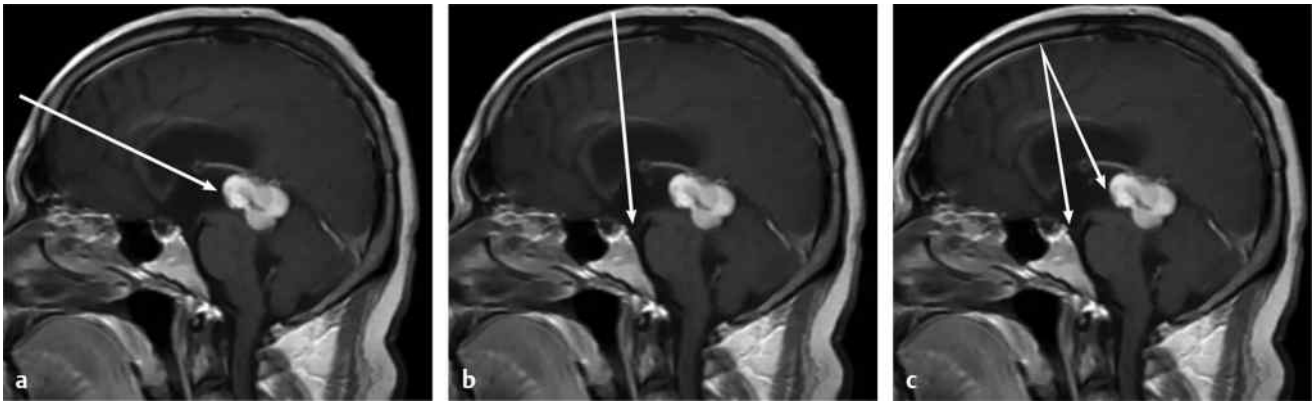


Fig. 24.2 Important trajectories in the treatment of pineal pathology. (a) The ideal transventricular route to the pineal region is from a frontal entry point (*arrow*), back through the center of the foramen of Monro to the pineal region. **(b)** The ideal route to perform an endoscopic third ventriculostomy is a line drawn back to the surface from the tuber cinereum backward (*arrow*), up through the foramen of Monro to the surface. **(c)** If the foramen of Monro is large enough and the tumor is anterior enough, it may be possible to do both procedures with a rigid endoscope through an intermediate trajectory. If so, both procedures should be performed with care to prevent anterior damage to the fornix and posterior damage to the thalamus.

tumor or to treat it with more conservative measures. Within the spectrum of PPT, pineocytomas are slow-growing tumors classified as WHO II lesions and they constitute 30 to 60% of PPT. Gross total resection of these tumors can be potentially curative. Since this patient was young and otherwise healthy, and there was a significant chance that his tumor was a pineocytoma, the decision for surgical removal was made with an aim on total resection.

This patient's tumor was in the pineal gland. It sat above the cerebellar vermis and tectal plate, behind the third ventricle, and anterior to the tentorial incisura in the midline (**Fig. 24.3**). Most importantly for the surgical approach, it was located below the internal cerebral veins and the great vein of Galen (**Fig. 24.4**). The basal veins of Rosenthal, as they join the internal cerebral veins to flow into the vein of Galen, are more lateral and, as such, they are less likely to be entangled by tumors of the region unless they are quite large. These deep venous structures were patent in our patient and must be preserved to ensure a successful clinical outcome. The only cranial nerve relevant to the area is the trochlear nerve. It usually courses forward below and lateral to the pineal gland.

Options for Approach

Although several surgical approaches are feasible, the two approaches to the pineal region most frequently utilized are the supracerebellar infratentorial approach and the occipital interhemispheric, transtentorial approach (**Fig. 24.5**). For PPTs, the supracerebellar infratentorial approach offers several natural advantages. As PPTs almost uniformly displace the vein of Galen and the other deep veins superiorly, these veins are shielded from the majority of the surgical dissection with the infratentorial supracerebellar approach. This approach, when performed in the midline, provides equal access to tumor components on either side. This is especially important when the tumor is wide in the coronal and axial plans.

There are of course disadvantages to the supracerebellar infratentorial approach as well. The difficulty of the approach is directly proportional to the steepness of the tentorium. There is currently no agreement in the literature for measuring the "angle of the tentorium," let alone defining the "steepness." In our own study, we chose to define the angle as measured against a line perpendicular to the floor of the fourth ventricle and found that it ranges from 45 to 75 degrees in the general population (**Fig. 24.6**). If a patient has a tentorial angle beyond 60 degrees, the supracerebellar infratentorial approach is extremely challenging, as it necessitates a long, steep "climb" along the undersurface of the tentorium. Flexing the cervical spine and using a sitting position can mitigate this challenge, especially in a thin patient with a supple neck. However, for a large patient with a thick neck, the supracerebellar infratentorial approach would be extremely difficult if the tentorium angle is above 60 degrees.

This weakness of the supracerebellar infratentorial approach coincides with the strength of the occipital interhemispheric, transtentorial approach. With the latter, a steep tentorial angle actually makes the operation easier as it brings the operative target closer to the surgeon. An example of a steep tentorial angle is shown in **Fig. 24.6**, and this patient's hemangioblastoma was resected via the occipital interhemispheric, transtentorial approach. This approach is done with the patient in the lateral position, and the ipsilateral side down, so that gravity can help move the occipital lobe away from the operative field. However, even with this, there is still significant risk of occipital lobe damage and therefore approach-related visual complications. The disadvantages of the occipital interhemispheric, transtentorial approach are that the approach is unilateral, and it comes from superior-to-inferior direction. As mentioned before, most PPTs displace the deep veins superiorly. Therefore, the occipital interhemispheric, transtentorial approach places the deep veins between the surgeon and target. Furthermore, if the PPT is big, the component of the tumor contralateral to the approach can be difficult to visualize as it may be hidden by the falx cerebri within the interhemispheric corridor. For these reasons, the supracerebellar infratentorial is generally preferred over the occipital interhemispheric, transtentorial approach for PPTs.

There is a rarely used third option to approach tumors of the pineal region, the parietal interhemispheric transcallosal approach. Compared to the more popular occipital interhemispheric option that reaches the target from a slightly

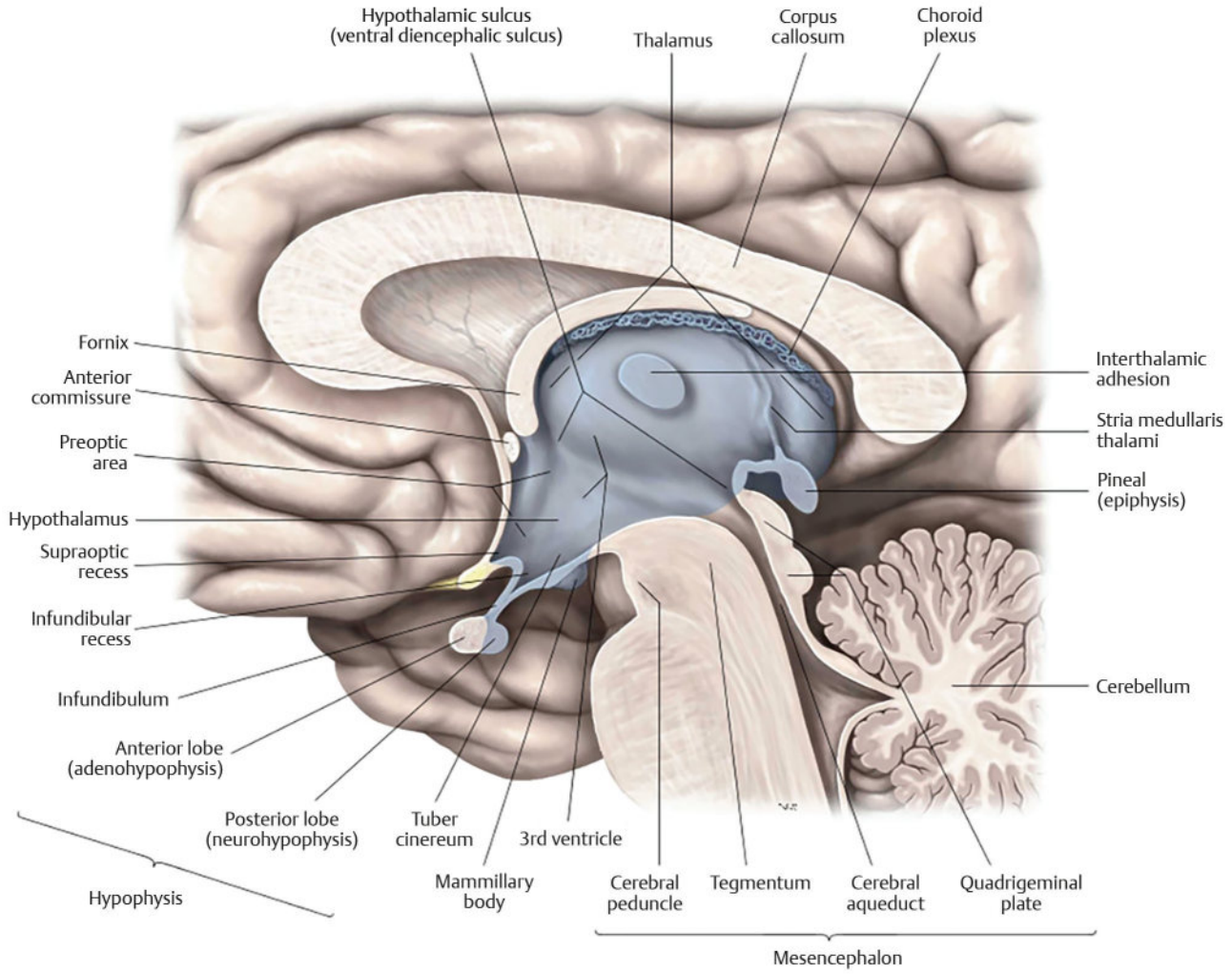


Fig. 24.3 Anatomical illustration. Sagittal diagram shows the pineal gland and adjacent structures. (Reproduced from Thieme Atlas of Anatomy, Neck and Internal Organs. Thieme; 2006. Illustration by Karl Wesker.)

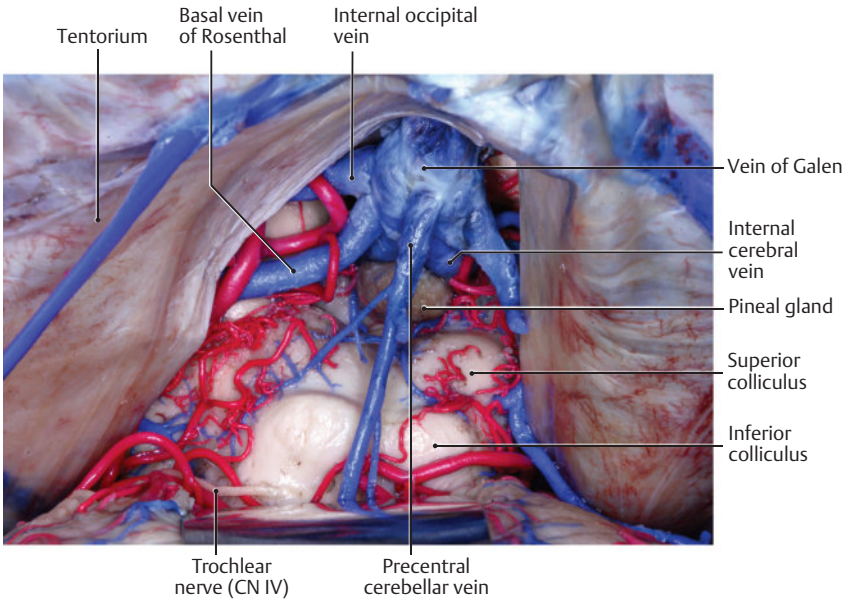


Fig. 24.4 Cadaveric dissection. Infratentorial dorsal view of the arteries and veins of the midbrain and pineal region. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st Ed. Thieme; 2017.)

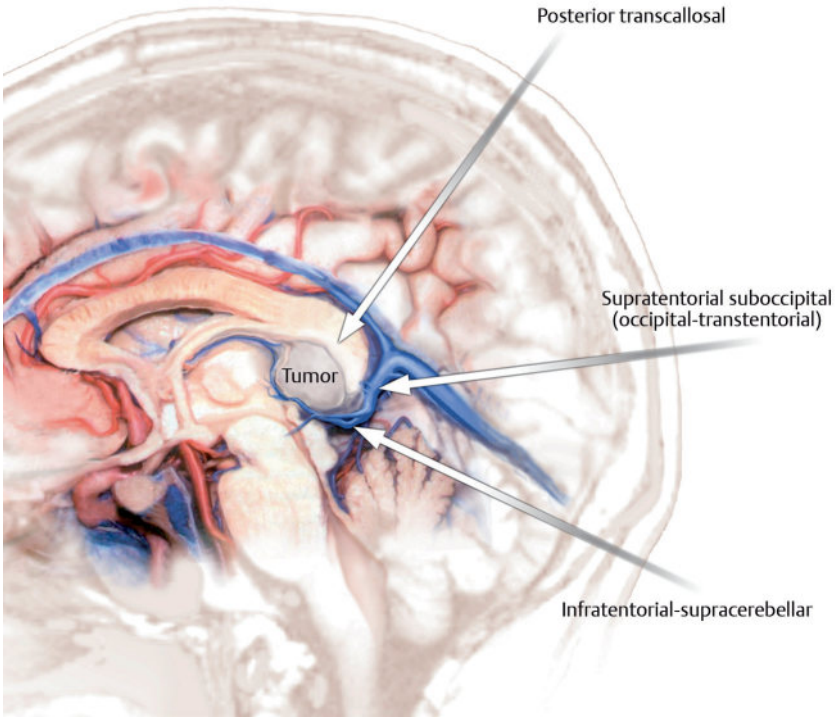


Fig. 24.5 Commonly utilized approach to the pineal region. (Reproduced from Nader R, Sabbagh A, eds. Neurosurgery Case Review: Questions and Answers. Thieme; 2010.)

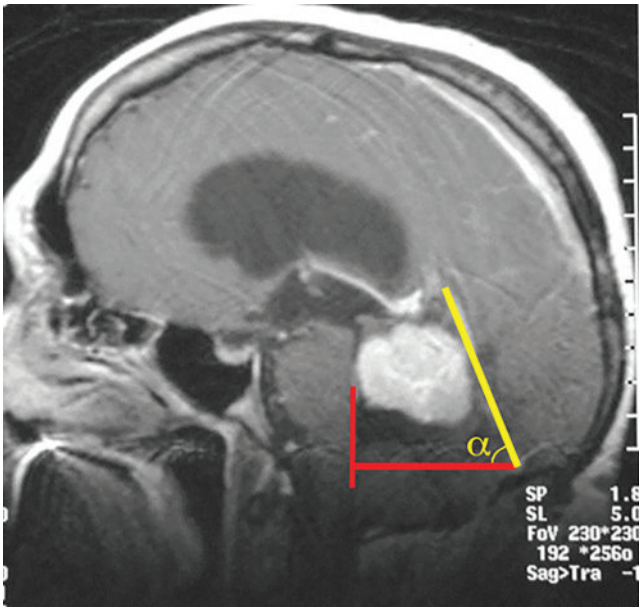


Fig. 24.6 Sagittal MRI with gadolinium of a different patient. This shows a uniformly enhancing tumor of the upper vermis. Notice the steep tentorial angle, α, as measured against a reference line perpendicular to the floor of the fourth ventricle. This, combined with the patient's large bony habitus, would have made an infratentorial supracerebellar approach in the sitting position very difficult. This hemangioblastoma was resected via an occipital interhemispheric transtentorial approach.

posterior-to-anterior trajectory, this approach reaches the target in a superior-to-inferior manner. One reason this option has gone out of favor is concerns with the interhemispheric bridging veins. Whereas in the occipital region, bridging veins draining into the superior sagittal sinus are quite sparse, going into the interhemispheric sulcus in the parietal region risks damage to the numerous bridging veins, especially the vein of Trolard. Furthermore, the risk of a disconnection syndrome resulting from incision in the posterior body of the corpus callosum is ill-defined. Thus, for these reasons, unless the pineal tumor is unusually anterior in its position, this approach is seldom used.

Questions

1. The precentral vein of the cerebellum is almost always blocking the approach to the pineal region during a supracerebellar infratentorial approach. How is this vein usually managed during this operation?
2. What is the most significant risk to the patient in a sitting position and what precautions must one take?
3. Is the supracerebellar infratentorial approach or occipital interhemispheric, transtentorial approach better for this patient?

■ Description of the Technique

Our patient was thin and had a long, supple neck. Furthermore, by the criteria described above, his tentorial angle was shallow at 45 degrees. Both the CTV and his MRI showed that the tumor clearly displaced the deep veins superiorly. For all those reasons, a supracerebellar infratentorial approach was chosen for this operation. Although several patient positions, including lateral and prone, are used for this operation, the preferred position is sitting. Gravity minimizes pooling of blood in the middle of the surgical field and mobilizes the cerebellum away from the path of the approach. Precordial Doppler and end-tidal CO_2 were both carefully monitored for indications of air embolism. A central venous catheter is also essential. In the event of an air embolism, the catheter can potentially remove the air (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: supracerebellar, infratentorial

Craniotomy: suboccipital

Modifiers: none

The patient was elevated from a supine position to the final sitting position by maneuvering the operating bed in a stepwise fashion. This must be performed gradually to avoid negatively affecting the patient's systemic blood pressure. In the final position, the head was affixed to a Mayfield head holder, and the cervical spine was flexed to bring the tentorium as close to parallel to the floor as possible. However, at the final flexed position, two fingerbreadths of space must be maintained between the patient's chin and manubrium to prevent elevation of airway pressure and compromising venous return.

An incision was made in the midline from 2 cm above the inion to the midcervical spine. The soft tissue was dissected laterally to expose the suboccipital region from the inion down to the foramen magnum. Although the incision was made down to the cervical spine for the purpose of exposure, the muscular attachments to the lamina were preserved. Two burr holes were made on either side of the midline just below the level of the torcula and transverse sinuses. Intraoperative navigation can be helpful in the placement of these burr holes. The craniotomy opening was made 2 cm from either side of the midline, from the level of the burr holes down to just above the foramen magnum. Opening the foramen magnum is usually unnecessary for this approach. After the bone flap was elevated, the bone overlying the torcula and transverse sinuses on both sides was burred away superficially, and rongeurs were used to carefully remove the final rim of bone covering these venous structures. Exposure of the inferior half of the transverse sinus is usually sufficient for the supracerebellar infratentorial approach (see **Operative Setup**).

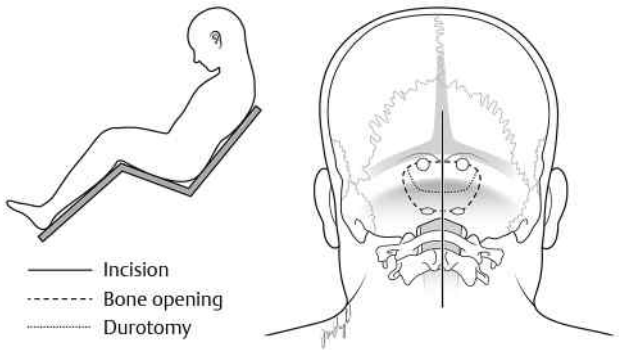
Operative Setup

Position: sitting

Incision: midline, inion to mid cervical spine

Bone Opening: midline suboccipital, approximately 2 square

Durotomy: "U" based on transverse sinus, reflected superiorly



The dura was opened in the shape of a "U," based on the transverse sinus. Opening in the midline supracerebellar infratentorial corridor then proceeded with careful dissection of any arachnoid adhesion encountered between the tentorium and the superior surface of the cerebellum. Bridging veins in the midline were coagulated and incised but similar veins existing laterally were preserved. The dorsal surface of the cerebellum was protected with cottonoids, and retractor blades were placed on top simply to secure the cottonoids and protect the cerebellum from the in-and-out surgical motions throughout the procedure, and not for traction purposes (**Fig. 24.7a**).

As the dissection approached the target, the usual arachnoid "curtain" was encountered (**Fig. 24.8a**). This veil separates the superficial surgical corridor from the quadrigeminal cistern and thus the whole pineal region and it must be sharply opened for the operation to continue (**Fig. 24.8b**). Once this was accomplished, the precentral vein of the cerebellum was identified as it traveled from the superior vermis to the vein of Galen. It was coagulated and safely divided.

The posterior capsule of the tumor was now exposed. It appeared grayish, with vessels draped on it superficially (**Fig. 24.8b**). The capsule was cauterized with the bipolar and then incised. The tumor was soft and quite vascular. It was debulked with suction, and as this process moved forward, the capsule was collapsed into the center (**Fig. 24.9**). At the end of the resection, the entire capsule was resected and the posterior third ventricle was widely opened. Both internal cerebral veins, as they entered the vein of Galen, were visualized and preserved. The vein of Galen itself was never visualized as it was superior to the surgical field and protected by surrounding arachnoid.

For closure, the dura was reapproximated with sutures and this closure was augmented with a dural substitute in an onlay manner. The craniotomy bone flap was replaced and the rest of the soft tissue closed. The skin was closed with a running nylon stitch to obtain watertight closure. The postoperative MRI showed a complete resection (**Fig. 24.10**).

Surgical Pearls

1. In choosing the supracerebellar infratentorial, one must anticipate the thick arachnoid veil that obscures the quadrigeminal cistern and the surgical target of the pineal region. Although sharp dissection may be counterintuitive, especially with multiple veins in close proximity behind it, it is nonetheless necessary. This veil is too thick for blunt dissection, and overaggressive blunt manipulation may in fact tear the precentral vein of the cerebellum directly behind it. Opening the veil sharply just lateral of the midline to avoid the precentral vein is the preferred maneuver.
2. Once behind the veil of arachnoid, the natural trajectory of the approach leads into the vein of Galen. For a tumor less than 2 cm in diameter, one must look inferiorly once the precentral vein is divided, as the top of the vermis would often "hide" the tumor. An adjustment in the retractor blade on the dorsal cerebellar surface is frequently necessary at this point to visualize the tumor.
3. Although the sitting position offers natural advantages for the supracerebellar infratentorial, this position may not be feasible with all patients. In particular, this position may not be safe for patients with a very high body mass index, and there exists a danger that such a patient may fall out of that position intraoperatively. Each anesthesia–nursing–surgery team must also be familiar and comfortable with the position. Although, blood can pool in the surgical field in the prone position, and there is no gravity-assisted mobilization of the cerebellum in the lateral position, both of these are possible alternatives if the sitting position is deemed unsafe.

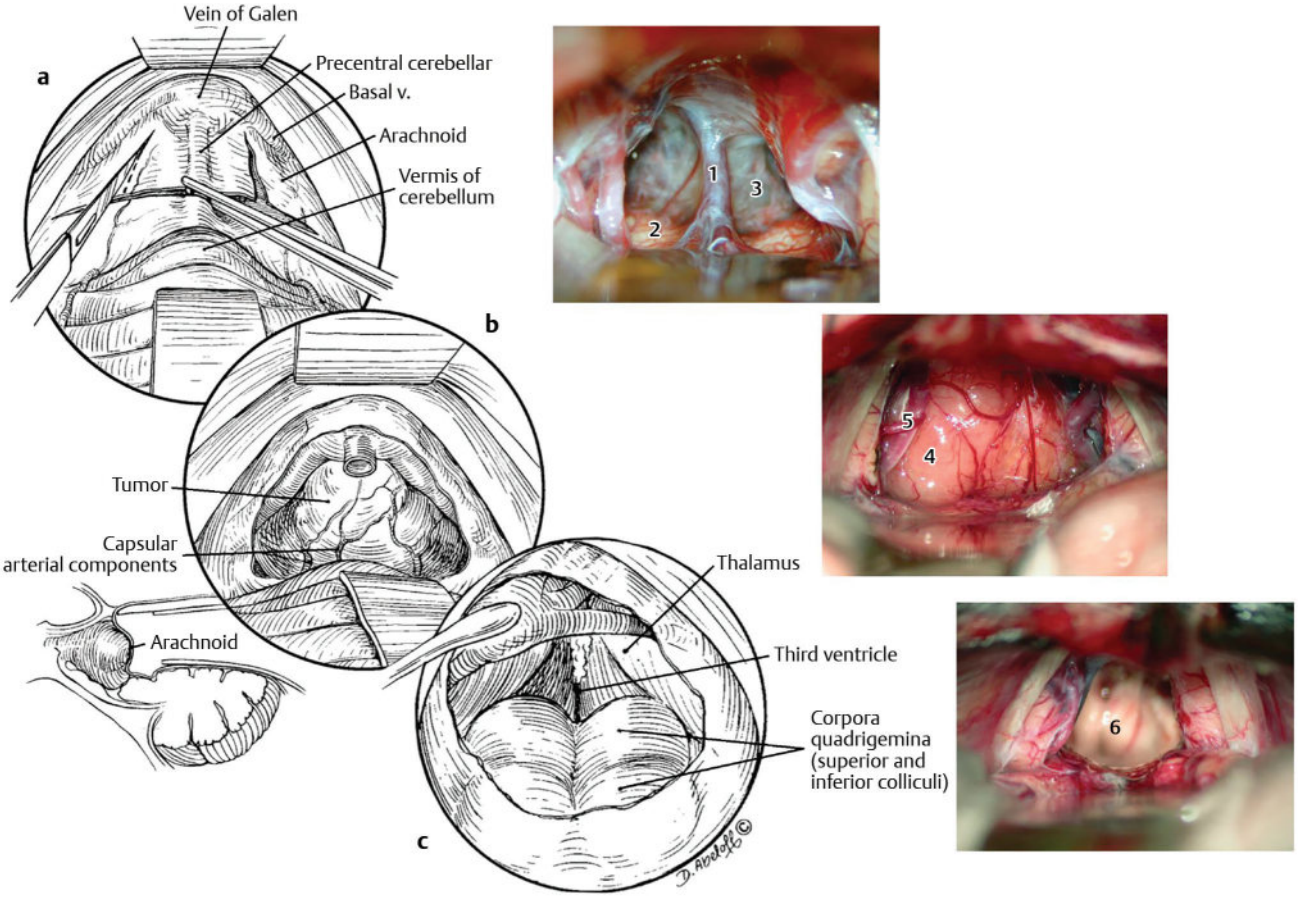


Fig. 24.7 The infratentorial supracerebellar approach. (a) Exposure of the precentral vein; 1, precentral vein; 2, cerebellum; 3, arachnoid. **(b)** Pineal tumor after precentral vein ligation and arachnoid dissection; 4, pineal tumor; 5, posterior choroidal artery. **(c)** View of the third ventricle after tumor excision; 6, third ventricle.

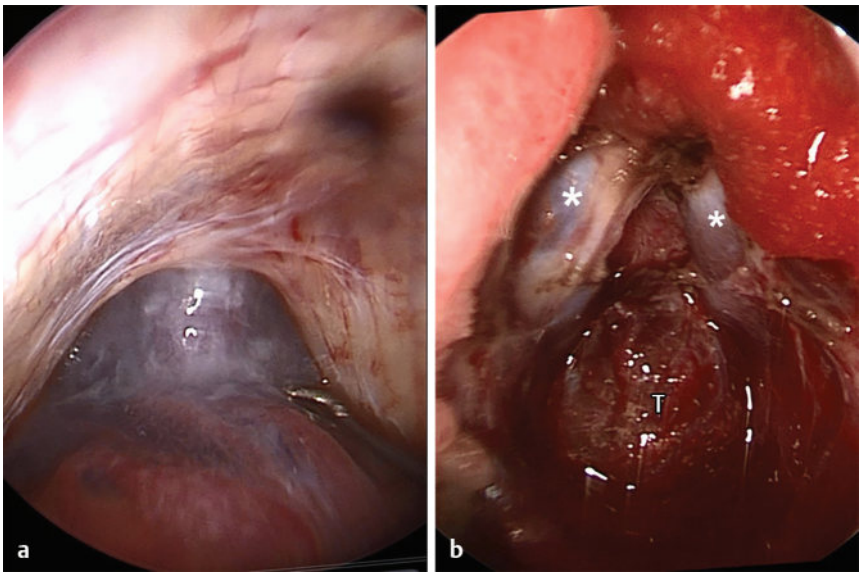


Fig. 24.8 Intraoperative views. (a) Surgical view under the tentorium, near the midline incisura. The arachnoid membrane blocking the surgical path to the pineal region was clearly seen. This member must be incised sharply. The precentral vein of the cerebellum can be seen coursing directly behind the membrane. **(b)** Surgical view of the pineal tumor (T). * denotes the distal internal cerebral veins as they join the vein of Galen.

■ Aftercare

The patient did well after his surgery with minor complaints of diplopia, as expected after sequela of surgery around the tectal plate. His head was elevated 30 degrees to minimize the chance of CSF leakage, and DVT prophylaxis was instituted soon after surgery. The patient was discharged 4 days after the resection. The histopathological diagnosis from the resection was once again "pineocytoma," but because of an unusually high MIB index (Ki-67), multiple opinions were obtained. The final

diagnosis from these consultations was "pineoblastoma," and thus he underwent craniospinal irradiation and chemotherapeutic treatments.

■ Possible Complications and Associated Management

Transient diplopia and upward problems are nearly universal after operations in the pineal area. For small tumors, these problems often resolve within days. Far more problematic are the risks of postoperative hemorrhage, CSF leak, and wound healing problems that can often complicate supracerebellar infratentorial surgery. However, the most dreaded complication may be venous air embolism related to the sitting position. Air embolism can cause hypotension and cardiac arrhythmias, and in severe cases, can be fatal. Because the operative area is above the level of the heart, negative pressure may exist in any of the large, exposed venous structures, and thus venous air embolism is prone to occur when performing the supracerebellar infratentorial in a sitting position. As previously discussed, end-tidal CO_2 and precordial Doppler should be carefully monitored throughout the procedure and a central venous catheter must be ready to remove the air when trapped. However, the most important prevention technique is awareness. Utmost care must be exercised to prevent opening into venous sinuses. But despite this, if a dural incision does violate what appears to be a sinus or large vein, and paradoxically there is *no bleeding*, the surgeon must know immediately that air is being sucked into the venous system. The opening must be covered promptly, and the surgeon must notify the anesthesiologist to prepare to remove the trapped air with the central venous catheter.

Postoperative hemorrhage in this operation does not only apply to the surgical cavity but also in remote supratentorial sites. The presumptive cause of these remote hemorrhages is once again the sitting position, as copious CSF is drained during the operation and traction related to gravity avulses small bridging veins over the supratentorial cortex. Fortunately, this complication is very rare. As with other posterior fossa operations, supracerebellar infratentorial can be complicated by CSF leakage. Meticulous dural closure, the generous use of dural substitute and sealant, and maintaining the head of bed above 30 degrees postoperatively can all reduce the incidence of leakage. If CSF leakage persists despite conventional interventions, such as lumbar drainage, one must remember that hydrocephalus can often follow surgery in the pineal region. This is especially true after removing large tumors in the region as postoperative debris inevitably runs into the posterior third ventricle. Early shunting may prevent downstream complications.

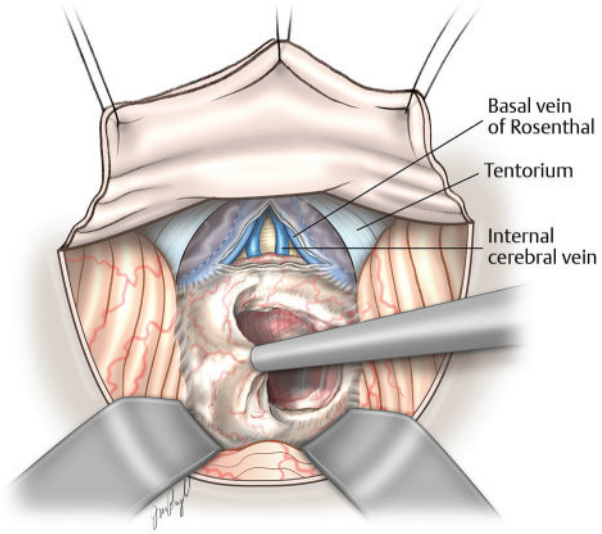


Fig. 24.9 Resection of the pineal tumor. As the tumor is debulked, the capsule is dissected from surrounding tissue and collapsed into the center.

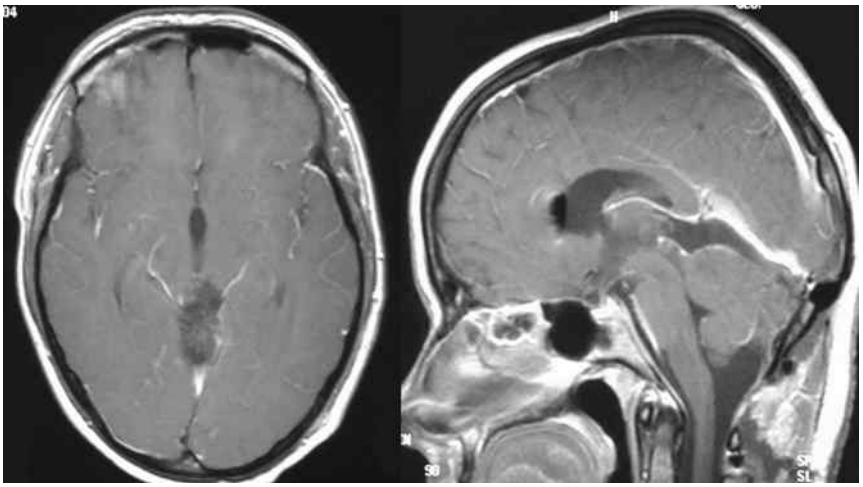


Fig. 24.10 Axial and sagittal MRI with gadolinium. Postoperative images showed a complete resection of the pineal tumor. The final histopathological diagnosis was "pineoblastoma."

Perspective

Michaela Lee and Peter Nakaji

Introduction

Multiple surgical options are available for addressing lesions in the pineal region, and several of these have been described in the previous section. Depending on the tumor characteristics and the anatomy of the individual patient, one approach may be more favorable than another. Ultimately, however, the option that the surgeon is most comfortable in using is often the best selection. As Charles Drake has often been quoted as saying, "Much of the merit of an approach is a matter of surgical experience."

For the vast majority of pineal region tumors, we favor the supracerebellar infratentorial approach, albeit with a few modifications. Other approaches, such as the occipital-transtentorial and the posterior interhemispheric, require substantial retraction of the occipital lobes in order for the pineal region to be visualized adequately, and doing so unnecessarily places the visual cortex at risk of injury. The senior author (PN) prefers a paramedian endoscopic supracerebellar infratentorial approach with the patient in a sitting-slouch position. The traditional approach, as described in the first section of this chapter, involves a long incision from the inion to the upper cervical spine with a generous craniotomy to expose the torcula and transverse sinus, and it often requires retraction on the cerebellum to provide sufficient illumination of the long surgical corridor under the microscope. We have found that having such a wide exposure superficially does not significantly improve surgical visualization, and it can increase the risk of exposure-related injury to the veins, sinus, torcula, and parenchyma. Instead, utilizing the endoscope with a small incision and a keyhole craniotomy provides a panoramic and well-illuminated view without the use of retractors. This technique provides a notable contrast to the experience with the microscope for this approach, which can sometimes give the surgeon an impression of working at the end of a long, dark tunnel.

Additionally, using a paramedian approach allows for better visualization of the pineal region because the view is unobstructed by bridging veins and the vermis (**Fig. 24.11**). Although the precentral vein can usually be safely divided, doing so is not necessary with a paramedian approach. Furthermore, the "steepness" of the tentorium might determine whether the midline supracerebellar infratentorial approach is feasible; it does not play a crucial role in the paramedian trajectory. This is because the steepness of the tentorium is maximal at the center and it gradually lessens as the tentorium extends out laterally. We generally prefer a left-sided craniotomy when the surgeon is right-handed so that the endoscope can be placed on the outer corner opposite the dominant hand to facilitate comfort and flexibility.

In the sitting position, the sagging of the cerebellum away from the tentorium (i.e., via gravity retraction) opens a natural infratentorial corridor. In addition, the tumor falls into view and the planes are better seen because of natural tension, thereby abolishing any need for fixed retractors. Pineal tumors generally hang down off the deep vein complex, which includes the internal cerebral veins anteriorly, traveling above the tumor in the posterior part of the velum interpositum to join the paired basal veins of Rosenthal, and the internal occipital veins, travelling laterally, to form the vein of Galen. Gravity also minimizes venous pressure, which translates to a more relaxed cerebellum and less blood loss, while whatever bleeding that does occur drains along with CSF away from the surgical field.

It is important to note that the sitting position is not feasible for all patients, particularly those with a patent foramen ovale, which conveys an increased risk of air embolism, and those with a very high body mass index because of the physical constraints of fitting such patients into the sitting-position frame. Aside from this small patient population, however, most patients can be placed in this position, so advantageous for pineal surgery. Alternatively, the endoscopic paramedian supracerebellar infratentorial approach can also be performed in a park-bench (or three-quarter prone) position.

The counterargument against endoscopic approaches mainly centers on the surgeon's unfamiliarity with endoscopes or the anesthesiologist's discomfort with patients in the sitting-slouch position. Early in their surgical careers, surgeons

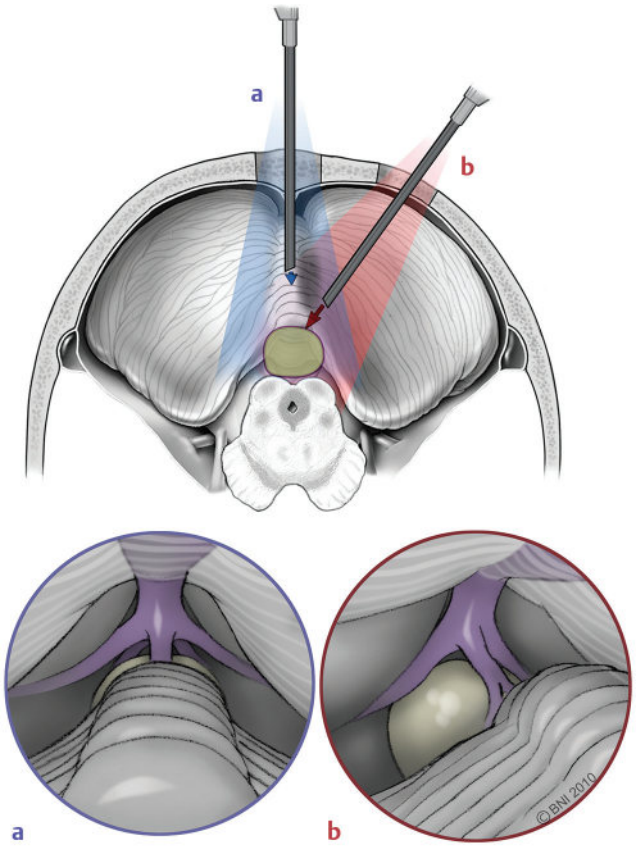


Fig. 24.11 Approach options. Illustration demonstrating **(a)** the midline trajectory and **(b)** the paramedian trajectory through the supracerebellar infratentorial space. The midline trajectory can be obstructed by the vermis and the bridging veins, whereas the paramedian trajectory provides a less obstructed view. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

may experience frustration with working in a narrow space and maneuvering instruments around neurovascular structures, for there is a great deal of critical anatomy that must be respected, regardless of the approach that is used. However, once the steep learning curve is past, the endoscope becomes a useful tool in the surgeon's armamentarium. It is the senior author's (PN) experience that the subtype of pathology (e.g., how large or calcified or vascular a lesion may be) becomes less relevant with increased surgical experience. As such, the paramedian supracerebellar infratentorial approach becomes more intrinsically favorable with the endoscope and allows for a simpler approach with lower impact on the patient.

■ Case Presentation

A 24-year-old woman was referred to our clinic for resection of a pineal mass. She had initially presented to a community hospital 2 months earlier with complaints of headaches, double vision, and syncopal episodes. At that time, she underwent a right frontal ETV and biopsy of the pineal mass that was complicated by a postoperative acute subdural hematoma requiring a large craniotomy for evacuation. Histopathological analysis identified the lesion as a WHO grade II PPT of intermediate differentiation. When we first saw the patient in our clinic, she was still recovering from mild left-sided weakness with some residual left grip weakness, and she complained of persistent headaches and near-syncopal episodes with rapid changes in position. The remainder of her neurological examination was unremarkable. She underwent a preoperative echocardiogram with bubble study to evaluate for a patent foramen ovale for which findings proved to be negative.

■ Anatomical and Therapeutic Considerations

MRI demonstrated a large (3 × 2 × 2 cm) avidly enhancing mass in the pineal region that displaced the tectum posteriorly (**Fig. 24.12**). The mass was mainly in the posterior third of the third ventricle, and it was easy to appreciate that the tumor enlarged the natural corridor just above the cerebellum. Given the young age of the patient and the pathologic findings from the biopsy, we believed that gross total resection was indicated. Some may consider the straight sinus too steep for a supracerebellar infratentorial approach, but as we pointed out previously, this is not a hindrance when using a paramedian trajectory. Therefore, we recommended a paramedian endoscopic supracerebellar infratentorial approach with a keyhole craniotomy for resection of this mass.

■ Description of the Technique

A lumbar drain was inserted preoperatively. Precordial Doppler probes and a central line were placed with the patient under

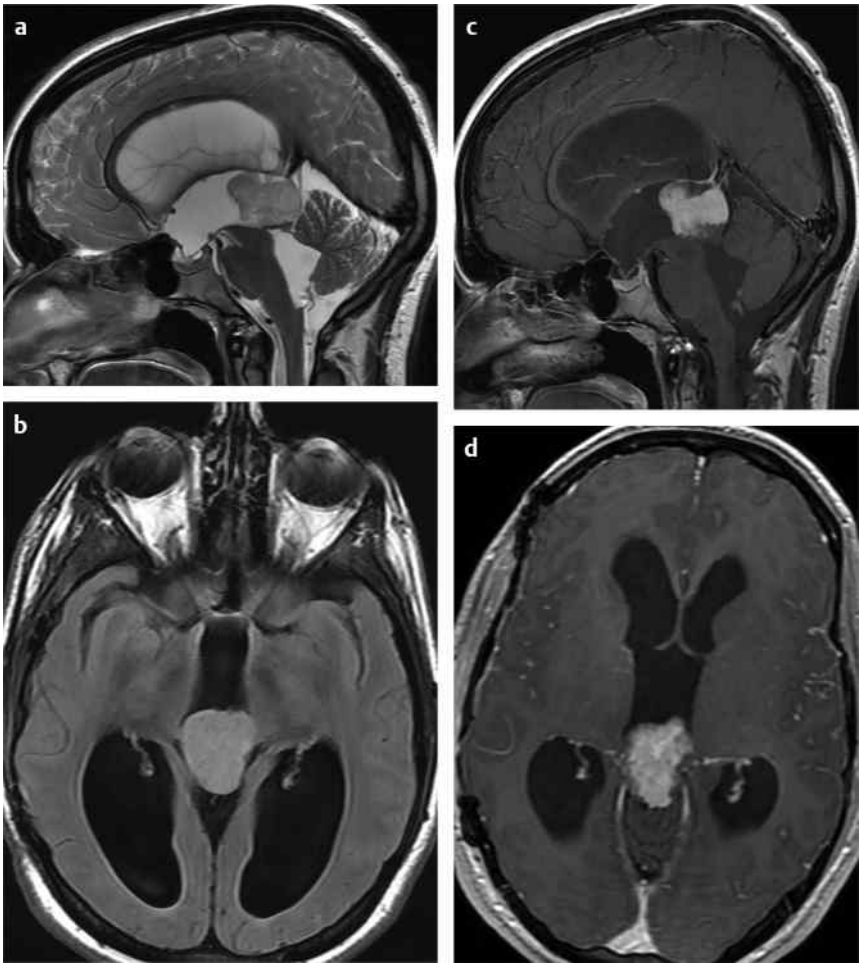


Fig. 24.12 Preoperative images. (a) Sagittal T2-weighted, **(b)** axial fluid-attenuated inversion recovery, **(c)** sagittal T1-weighted with contrast, and **(d)** axial T1-weighted with contrast MRIs demonstrated an avidly enhancing mass in the pineal region at the posterior third of the third ventricle, causing mass effect and displacing the tectum posteriorly with associated ventriculomegaly. A right-sided craniotomy from previous evacuation of a subdural hematoma was also evident. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

anesthesia to monitor for any air embolism. She was placed in a sitting-slouch position with a Mayfield head holder. Image guidance was used to localize the torcula and the transverse sinus based on the preoperative MRI. A 4-cm paramedian vertical incision was made, 2 to 2.5 cm off midline, starting slightly above the level of the transverse sinus and extending downward. A keyhole craniotomy (2 × 2 cm, **Fig. 24.13a**) was then performed to expose the dura from the lower part of the transverse sinus and the upper cerebellum. The dura was opened in an inverted U-shaped fashion based on the transverse sinus. With gravity and aspiration of CSF, the cerebellum relaxed nicely to reveal a wide surgical corridor (**Fig. 24.13b**). The cerebellum was protected with long Telfa (Medtronic, plc, Dublin, Ireland) pads and the zero-degree endoscope was advanced over this surface. Retractor blades were not necessary and all bridging veins and the precentral cerebellar vein were preserved. A large pineal tumor could be seen just lateral to the basal vein of Rosenthal (**Fig. 24.13c**). At this point, the tumor was debulked (**Fig. 24.13d**) using a two-handed microsurgical technique while the assistant held the endoscope in the upper left-hand corner of the craniotomy. While maneuvering in and out of the operative field, the endoscope always followed the instruments to ensure that the tips of the instruments were seen. Side-to-side swings with any tool were avoided to prevent iatrogenic injuries to structures not visualized by the endoscope.

A specimen was obtained for both frozen and permanent pathologic evaluation. The rest of the tumor was then removed using meticulous circumferential dissection. A 30-degree endoscope was brought into the surgical field at the end of the procedure to enable the surgeon to visualize the third ventricle and confirm resection of the entire tumor (**Fig. 24.13e**). After copious irrigation and achievement of hemostasis, a DuraGen onlay (Integra LifeSciences Corp., Plainsboro, NJ) was placed under the dura and dural replacement material was used to construct a watertight closure using 4–0 nylon sutures. The bone flap was replaced with titanium plates, and the scalp was closed with interrupted polyglactin stitches and running nylon. The lumbar drain, draining 5 mL/h, was left in place for 1 day postoperatively.

■ Aftercare

During the immediate postoperative period, the patient had a mild left pronator drift that resolved by postoperative day 2. Her pupils were reactive and equal, extraocular muscles were intact with conjugate gaze, and she had no diplopia. MRI demonstrated gross total resection of the tumor. Pathologic results were consistent with a PPT of intermediate differentiation. On postoperative day 8, the patient developed increased and symptomatic hydrocephalus despite the previous ETV, and she required placement of a left parieto-occipital ventriculoperitoneal shunt. A month after her surgery, she had a seizure and was brought to the emergency department; a CT of the head showed a left chronic subdural hematoma. The hematoma was evacuated using burr holes, and the patient recovered well. Three months after the index operation, she underwent stereotactic radiosurgery (Gamma Knife, Elekta AB, Stockholm, Sweden) to the surgical bed. Overall, she has done quite well and has had no neurological deficits. At her most recent follow-up

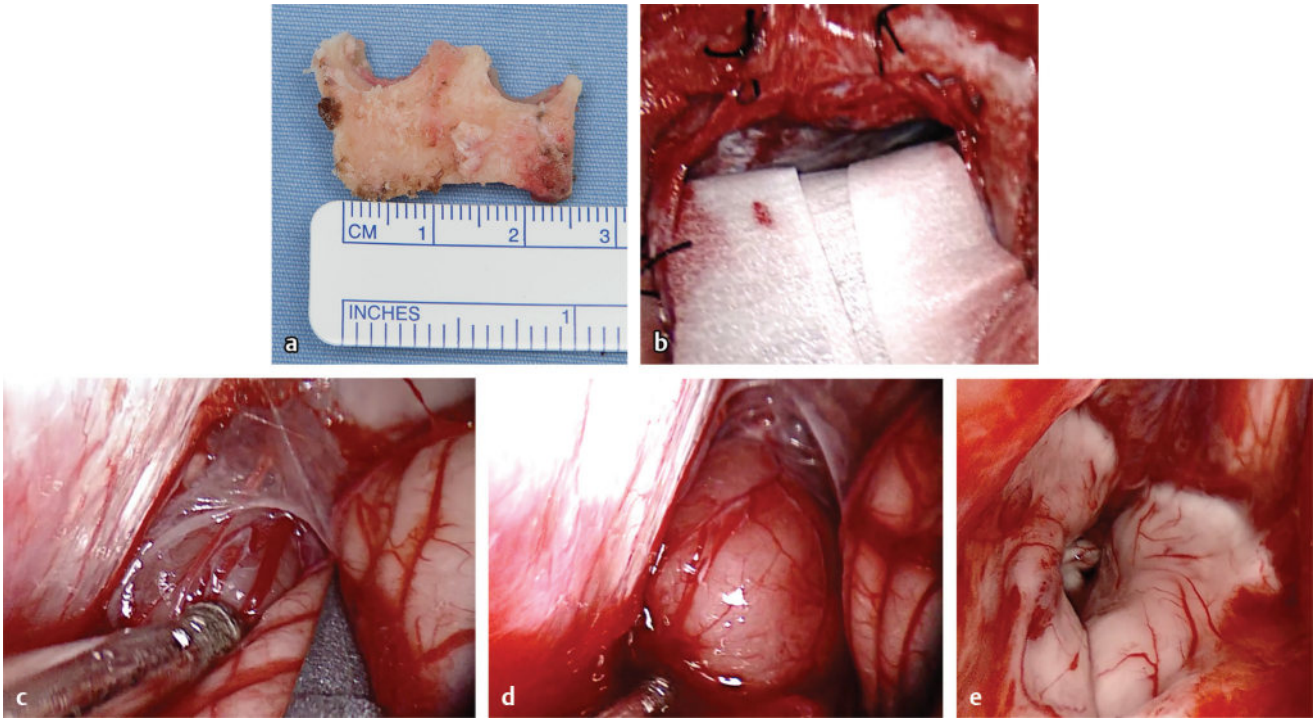


Fig. 24.13 Intraoperative views. Images from resection of a pineal mass using a paramedian endoscopic supracerebellar infratentorial approach. **(a)** Bone from a keyhole craniotomy measuring about 2 × 2 cm. **(b)** Dural opening and surgical corridor. **(c)** Arachnoid covering the tumor is opened and the basal vein of Rosenthal can be seen to the right of the tumor. **(d)** Intraoperative view of pineal tumor with tentorium on the left and cerebellum on the right. **(e)** Resection cavity after gross-total resection of the tumor. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

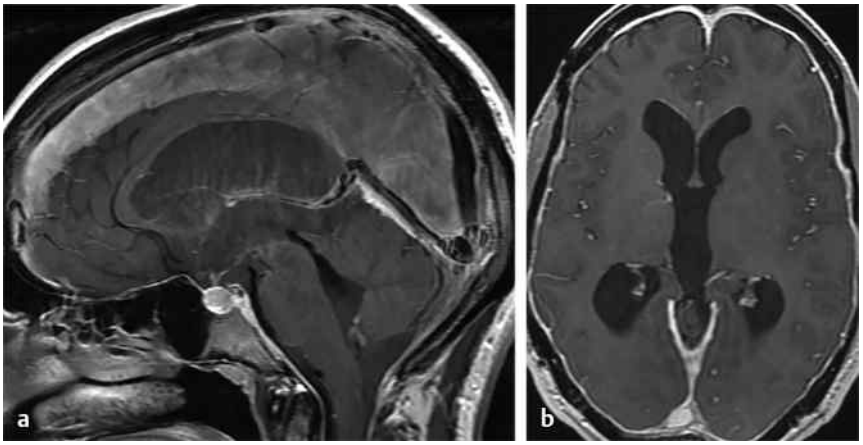


Fig. 24.14 Postoperative images. (a) Sagittal T1-weighted and **(b)** axial T1-weighted with contrast MRIs at 28 months postoperatively showed complete resection of pineal tumor with no evidence of recurrence. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

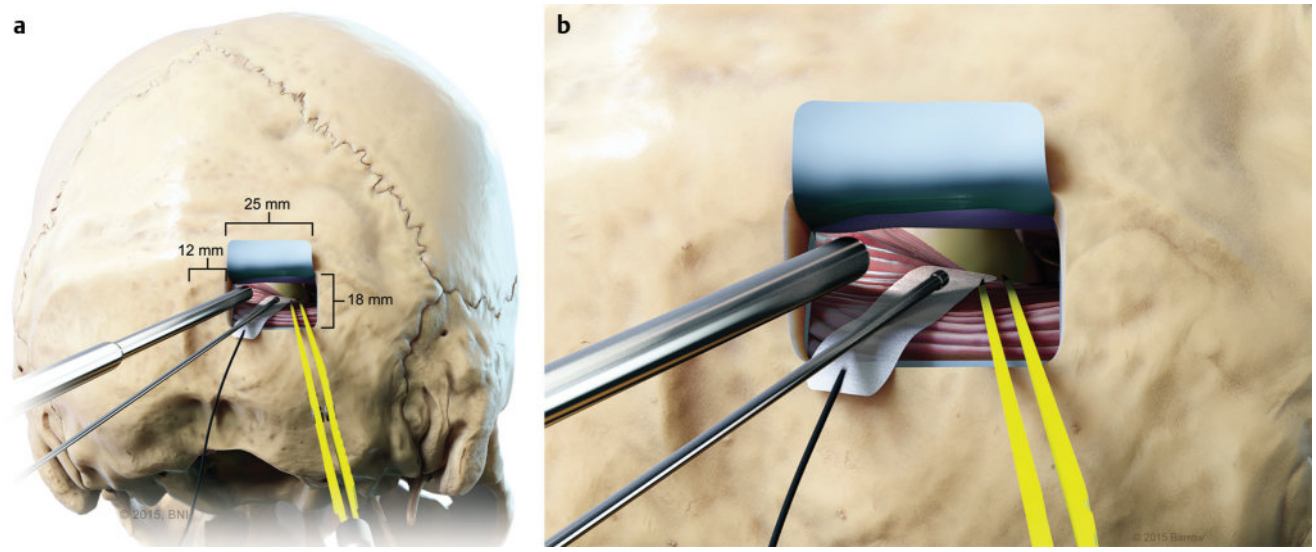


Fig. 24.15 Keyhole concept. (a) Illustration demonstrating the size of a minicraniotomy and the four-handed approach. **(b)** Illustration showing magnification of surgical corridor. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

MRI at 28 months postoperatively, there was no recurrence of tumor (**Fig. 24.14**).

■ Commentary

With the adoption of new technology and instruments, we must be open to creative and unconventional techniques, which may ultimately minimize the potential morbidities for our patients. One of the main goals in any surgery is to damage as little brain as possible, regardless of whether the tissue is placed at risk during the approach by retraction or direct surgical manipulation. In the case described in this section, the paramedian endoscopic supracerebellar infratentorial approach with a keyhole craniotomy minimized these risks, which exemplifies the advantages of using an endoscope and deviating from the traditional midline approach.

With the patient in a sitting position, the cerebellum will relax with the force of gravity, the aspiration of CSF, and the use of mannitol. A neuronavigation adapter can also be attached to the endoscope for better anatomical orientation, if necessary. Before the start of the operation, all precautions to prevent an air embolism should be made, and the surgeon should be familiar with managing any intraoperative air emboli. The natural corridor reveals itself almost effortlessly as one makes one's way toward the pineal region without unnecessary injury to veins, torcula, sinus, or cerebellum. Even with a much smaller craniotomy, the surgical corridor is wide enough for almost any resection and the field is well-illuminated with the endoscope (**Fig. 24.15**).

It is important to remember that the trajectory to the pineal region is not absolutely parallel to the tentorium, but rather is slightly tilted inferiorly to reach the thick white arachnoid membrane over the confluence of veins that enter the vein of Galen. We advocate the multihanded approach, with the assistant holding the endoscope while the surgeon operates with two hands using standard microsurgical tools. Once the thick arachnoid is sharply dissected, the precentral cerebellar vein is exposed (but not sacrificed) and the paramedian exposure allows access to both the ipsilateral and contralateral pineal region from one side. The more oblique angle gives a less-obstructed view not only of the pineal region but also of the splenium of corpus callosum by avoiding the obstruction from the culmen.

During dissection of the tumor, one must carefully preserve all the major veins, including the internal cerebral veins and the basal veins of Rosenthal, as well as exercise utmost

caution not to pull on the tectum and its associated microvasculature. A thorough tumor removal will reveal the posterior third ventricle. An angled-endoscope can be brought into the surgical field to visualize the anterior third ventricle and down to the proximal cerebral aqueduct; however, reaching those areas with surgical instruments is likely to impact the tectum negatively, because the angle is not conducive to working in these areas.

The paramedian endoscopic supracerebellar infratentorial approach is also very versatile. The superior surface of the cerebellar hemispheres flattens as one goes laterally from the midline. We have found that the more lateral the trajectory, the greater the degree of surgical freedom to access the pineal region, tectum, and posterior incisura. Hence, one can modify the technique as needed to achieve a lateral or extreme-lateral supracerebellar infratentorial approach. With the illumination and panoramic view of the endoscope, the approach can be further customized to a supracerebellar transtentorial approach to gain access to lesions of the tentorial incisura as anteriorly as the medial temporal lobe and as far posteriorly as the occipital lobe.

There are multiple passages to the pineal region. Although there is no one single "correct" approach and each approach must be customized to the patient, alternatives such as the paramedian endoscopic supracerebellar infratentorial approach can minimize potential morbidities. As neurosurgeons, we must actively pursue advancements in surgical approaches for overall improvement in all aspects of patient care and to ensure the continued development of our specialty. The learning curve is understandably steep, as it includes not only the development and refinement of endoscopic skills, but the acquisition of new anatomical understanding as one sees the structures from unconventional angles. But the approach presented here can be an excellent option, using only a small incision and keyhole craniotomy to provide a high-definition, well-illuminated view, while minimizing approach-related morbidity for patients.

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Part VIII

Tumors of the Posteroinferior Skull Base

25 Brainstem (Pontomesencephalic) *323*

26 Brainstem (Pontomedullary) *332*

27 Cerebellomedullary Fissure *342*

28 Torcula *352*

25 Brainstem (Pontomesencephalic)

Walter C. Jean

Keywords: brainstem tumor, pontomesencephalic, tentorium, vein of Galen, cavernous malformation

■ Case Presentation

A 69-year-old man presented with the chief complaint of dizziness, blurry vision, and perioral numbness for 2 months. His medical history is significant for melanoma, which was removed from his upper back about 8 years previously. He did not undergo further treatment, such as chemotherapy, for the melanoma after its removal. On examination, he had some patchy numbness to light touch on the face and his gait was unsteady. The remainder of his examination was unremarkable. The patient's magnetic resonance imaging (MRI) showed a cystic lesion in the pontomesencephalic junction with no visible solid component associated with it (**Fig. 25.1**).

Questions

1. What is the differential diagnosis based on the MRI finding and the patient's history?
2. Is there any family history question we need to ask to help us with the differential diagnosis?
3. Given the location of the lesion and his physical examination, what is the most rational management?

■ Diagnosis and Assessment

Within the differential diagnosis of brainstem-related lesions in adults, one can immediately rule out two of them by the MRI appearance: a neuroenteric cyst would show high-signal on T1 and a cavernous malformation would have a heterogeneous appearance with hemosiderin. Despite the lack of a solid component, a hemangioblastoma is still worth considering. It is therefore obligatory to ask the patient for his family history, and even though his family was free of von Hippel–Lindau disease, it does not completely rule out a hemangioblastoma. The most likely diagnosis for this patient, however, is a primary brainstem glioma and a metastasis. Given his history of melanoma, the latter diagnosis was favored.

The absence of a solid nodule made biopsy quite challenging, whereas the cyst made stand-alone radiation possibly ineffectual. The decision was therefore made to drain the cyst fluid via a stereotactic-guided aspiration, with the hope of a cytological diagnosis from the fluid sample, and a reduction of the overall volume of the mass to make radiation therapy feasible. The biopsy needle was advanced via a frontal burr hole through the cerebral peduncle into the cyst, and approximately 20 cc of yellowish fluid was removed. Unfortunately, the contents of the fluid did not yield a diagnosis, but with the collapse of the cyst wall, the patient underwent radiation therapy (5400 cGy over 6 weeks), with the presumptive diagnosis of metastatic tumor.

With that, the patient returned to his neurological baseline until precisely a year later, when he started to experience leg weakness, which led him to frequent falls. He also started to have swallowing difficulties. His physical examination at this time showed leg weakness, more on the right, and absent gag reflexes on both sides. While the patient's body imaging remains clear of any systemic cancer, the brain MRI taken at this point showed a recurrence of his cyst but this time it was associated with a significant solid component on the left side of the pons (**Fig. 25.2**).

As radiation therapy was no longer a viable option, the patient opted for surgical treatment to combat the expected symptomatic and radiographic progression of his tumor.

■ Anatomical and Therapeutic Considerations

With the development of a solid tumor next to the cyst, a combination procedure for biopsy of the tumor and repeated

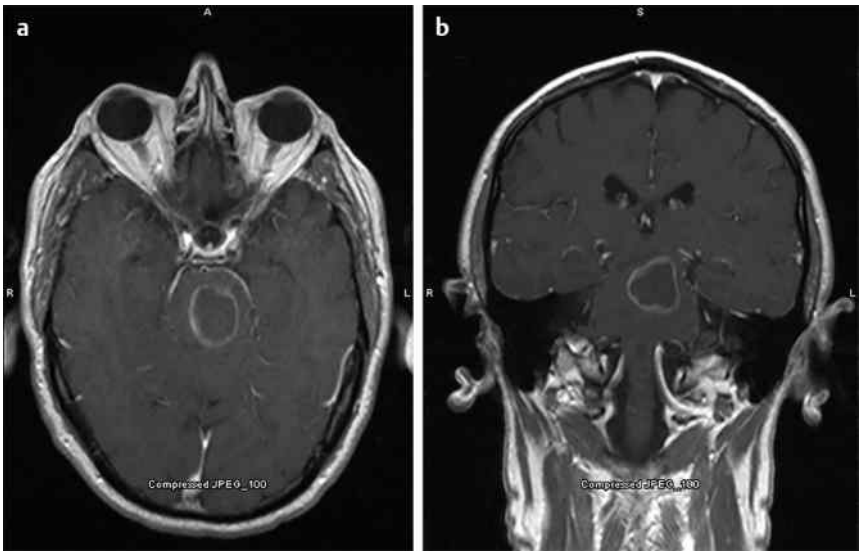


Fig. 25.1 MRI at first presentation. (a) Axial and **(b)** coronal images showing a cystic mass in the pons with no discernable solid portion.

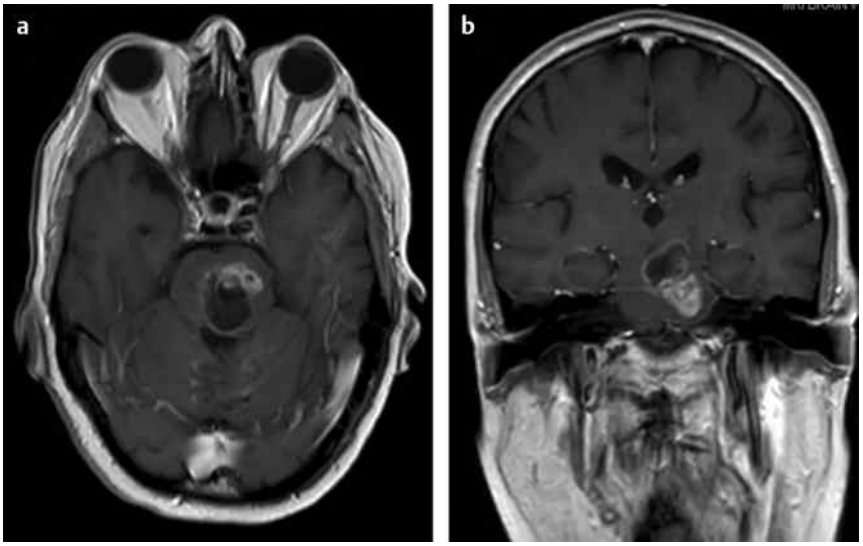


Fig. 25.2 MRI 1 year after radiotherapy. **(a)** Axial and **(b)** coronal images showing the cystic mass in the same patient one year after radiation treatment. At this point, the tumor had a solid component on the left side.

drainage of the cyst is a viable surgical option. Further treatment would then be dependent on the histopathological identity of the tumor. Several problems were readily apparent with this idea: as radiation was no longer a possibility, the cyst would likely reaccumulate unless the tumor were responsive with chemotherapy. Even though the patient's body imaging was not able to detect any systemic cancer, the prevailing thought remained that the tumor was likely metastatic in nature, and because few, if any, metastatic brain tumors can be controlled with chemotherapy alone, the idea of biopsy/drainage followed by chemotherapy had significant flaws. This option can only be considered "palliative."

The other option to consider was surgical removal of the entire cystic tumor. The success of this option would be highly dependent on not only the tumor type, but also the anatomical relationship between the tumor margin and the brainstem. Most of the outcomes data that exist on adult brainstem surgery pertains to cavernous malformations, for which there exists a definable demarcation between lesion and brainstem. The margin between tumor and brainstem in this patient is probably much less clear-cut, as any definable edge would likely have been obscured by the effects of the radiation rendered a year prior.

The fact that his symptoms were rapidly progressing, and he was quickly changing from a person, who was fully functional at work and in life, to a patient, who was dependent on his caregiver for all his needs, probably influenced this patient's decision. Posited between two options, both of which had significant flaws, he chose the more aggressive one, with the faint hope that under the "best case scenario," he might be able to maintain or regain his functional independence. He was also aware that any scenario other than the best would probably result in significant new deficits, and the possibility of needing both a tracheostomy and gastrostomy to maintain basic physiological functions.

This patient's tumor was located mainly in the pontomesencephalic junction. The solid component extended from the midline to the left in the pons, and the cystic portion extended as superiorly as the diencephalon. The solid mass reached closest to the surface inferior to the root entry zone of the left trigeminal nerve, whereas the cyst came closest to the brainstem surface just under the inferior colliculus in the midline under the frenulum veli. The exit of the trochlear nerves is just lateral to this area. The key to designing the operative approach is finding a way to enter the brainstem, in the safest way possible, to reach the tumor where it is near the surface, but the surgical corridor must provide sufficient exposure for both the solid component in the pons as well as the cystic part that extends superiorly.

Options for Approach

As previously mentioned, nearly all the literature written on adult brainstem surgery pertains to removal of cavernous malformations, and the concept of "safe entry zone" into the brainstem has emerged from this body of work. These zones are simply areas where there are *relatively* fewer motor fibers and cranial nerve nuclei, where incision into the brainstem would cause the least damage in theory. Even though logic would instruct us to attack the lesion where it is closest to the brainstem surface, a surgical approach designed simply to reach this point may necessitate an awkward or dangerous path to the center of the tumor (i.e., target). The entry point into the brainstem must be considered in conjunction with the safest trajectory to the target. As such, unlike the majority of other skull base tumors, for which the approach corridor is a one-step consideration (i.e., surgeon-to-target), for this brainstem tumor, the design of the approach is a two-step process: (1) surgeon-to-brainstem entry and (2) brainstem entry-to-target. We must first determine the safest point to enter the brainstem, and then design the proper exposure to reach this point.

Since our tumor is far from the ventral pontine surface, safe entry zones on that surface are out of consideration. An entry site in the pontomedullary sulcus can be reached via a far-lateral suboccipital approach (**Fig. 25.3a**), and superior fovea entry site, bordered by the triangle (superior cerebellar peduncle-vestibular area-sulcus limitans), can be reached by a telovelar approach (**Fig. 25.3b**), but both of these approaches would have difficulty reaching the superior midbrain component of our patient's tumor. Similarly, any surgical corridor with a straight superior–inferior or inferior–superior trajectory can be eliminated because of difficult reach at the extremes.

Logically, we must next consider the lateral approaches. A lateral entry point at the mid-pons level would reach the solid portion first, and the "peritrigeminal" safe entry zone would be suitable for this purpose (**Fig. 25.3**). Various surgeons have writ-

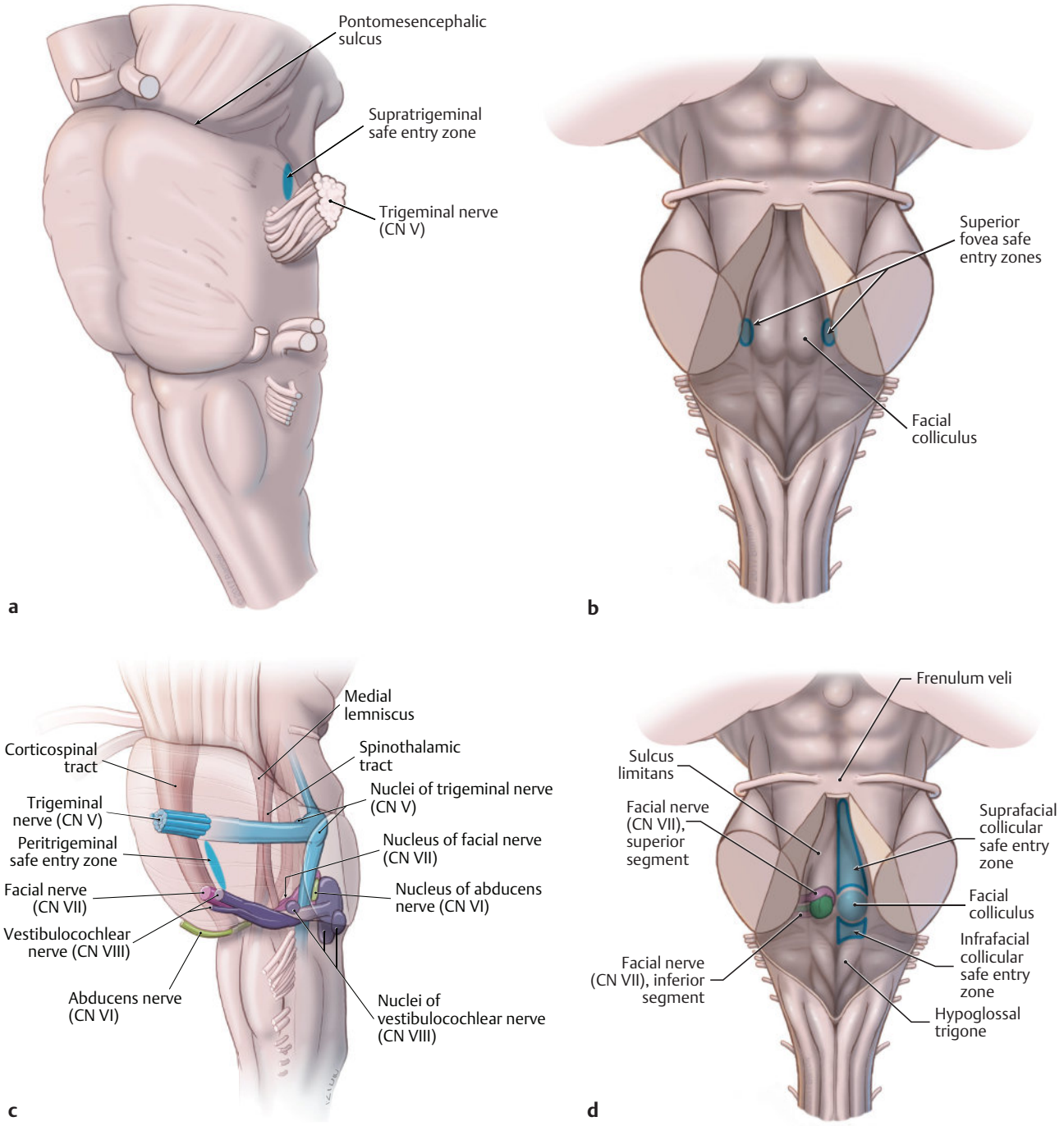


Fig. 25.3 Safe entry zones of the pons. (a) Ventrolateral view of the pons showing the pontomedullary sulcus and supratrigeminal safe entry zones of the brainstem. **(b)** Dorsal view of the pons with the superior fovea safe entry zone which is bordered by the superior cerebellar peduncle, the vestibular area, and sulcus limitans. **(c)** View of the peritrigeminal safe entry zone and **(d)** the infracollicular entry point that was chosen for our patient. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

ten about entry sites around the trigeminal root and given the sites various names. For example, Ferroli et al described using a retrosigmoid craniotomy to reach an "infratrigeminal" entry point just under the trigeminal root, whereas Zenonos et al used the subtemporal, transtentorial corridor, via a middle fossa craniotomy, to reach a supratrigeminal entry point just superior to it (**Fig. 25.3a**). The idea of using the subtemporal corridor with incision of the tentorium holds a lot of promise, as it would not only provide access to a safe entry zone near the trigeminal nerve, but also sufficient exposure for the resection of the entire lesion.

Another option is to attack the cyst first, and a reasonable safe entry zone to use would be the infracollicular area (**Fig. 25.3d**). As the name implies, this zone is between the lower edge of the inferior colliculus and the exit of the trochlear nerve. To reach this entry zone, the supracerebellar, infratentorial corridor must be considered, but the tentorium would restrict the surgeon's line of sight to an inferior–superior trajectory, and like the previous considerations with similar properties, this approach would have difficulty reaching the entire lesion. Alternatively, the infracollicular entry zone can be reached via the inter-

hemispheric transtentorial approach. The main disadvantage of this idea is that the deep veins, including the vein of Galen, are between the surgeon and the target. However, if one could safely dissect and protect these veins within the surgical corridor, this approach would provide excellent and direct exposure to the entry zone and the entire lesion.

Approach of Choice

There are significant advantages in this patient to approach the cyst first. The release of the cyst fluid always provides the surgeon unequivocal reassurance, and often, much-needed relief, that the navigation is on-track during a high-anxiety search-mission deep in eloquent brainstem tissue. Second, between the two components of our patient's tumor, the cyst is closer to the surface of the brainstem (in the infracollicular area) than the solid part (in the peritrigeminal area). And finally, a cyst is usually considered the "weak side" of a tumor more suitable for surgical approach because the cyst cavity itself provides excellent exposure to the entire tumor, as it is an empty space that can be converted into a "working room" for the surgeon.

For these reasons, the interhemispheric transtentorial approach was chosen to enter the brainstem in the infracollicular safe zone for the resection of our patient's tumor.

Questions

1. Which side would you approach this tumor from?
2. Given the choice of an interhemispheric approach, how would you position the patient for surgery?
3. What type of expectations would you set with the patient and his family regarding possible outcomes?

Description of the Technique

Because of the orientation of this particular tumor with the solid part projecting downward to the left, a contralateral approach from the right side was chosen to provide a straight line of sight for the surgeon. The patient was positioned in a lateral position with the right side down. The neck was placed in a completely neutral position without flexion or rotation to either side. The transverse sinus was approximated using external landmarks and neuronavigation. The planned incision was in the shape of the letter "U." The skin flap, overlying the right occipital lobe, was based laterally (see **The Three Approach Elements**).

After reflecting the skin, galea and pericranium laterally, neuronavigation was used to identify the transverse and superior sagittal sinus (SSS). Two burr holes were made in the midline on the SSS, with two additional ones more laterally. A rectangular bone flap (approximately 5 × 3 cm) was elevated with utmost care not to violate the SSS. The dura was opened with a "U" shape based laterally, away from the SSS (**Fig. 25.4a**).

As gravity-assisted retraction mobilized the ipsilateral occipital lobe laterally, dissection of the interhemispheric fissure proceeded with caution so as not to damage cortical veins (see **Operative Setup**). As predicted, no bridging veins from the

The Three Approach Elements

Corridor: interhemispheric

Craniotomy: occipital

Modifier: incision of the tentorium

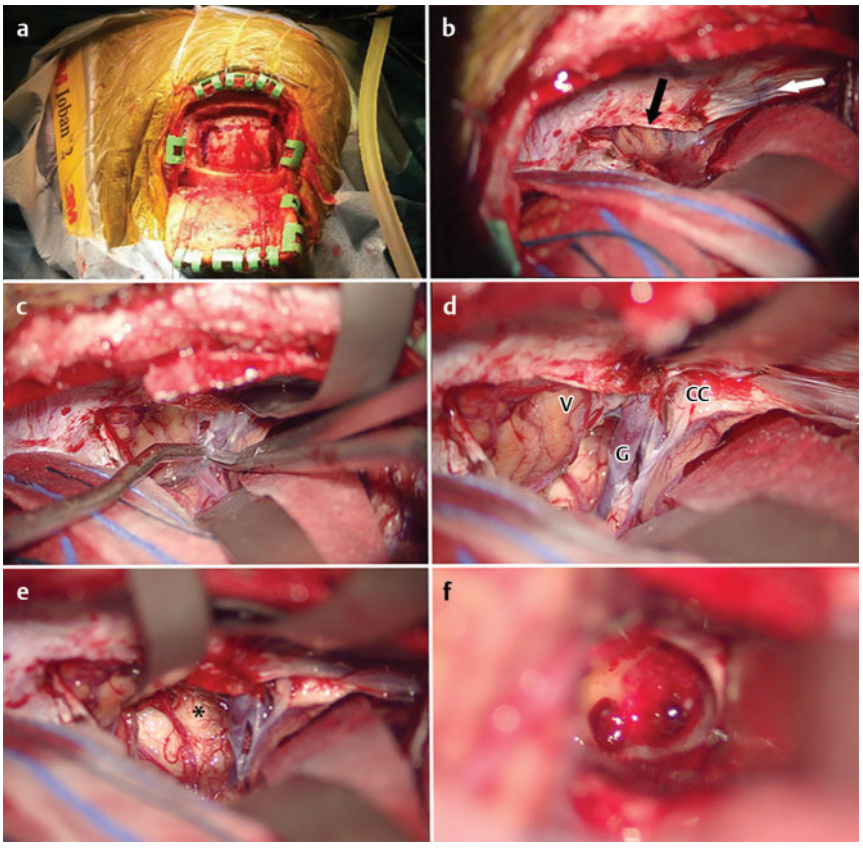


Fig. 25.4 Intraoperative images. **(a)** A right-sided occipital craniotomy was completed and the planned durotomy was marked in purple ink. The dural flap was based laterally and the medial cut was just to the right of and parallel to the superior sagittal sinus. **(b)** Gravity-assisted retraction has mobilized the right occipital lobe away, exposing the falx and tentorium. *Black arrow*, cut edge of the tentorium; *white arrow*, straight sinus. **(c)** The arachnoid covering the vein of Galen was dissected. **(d)** The vein of Galen (G) and the splenium of the corpus callosum (CC) are shown in this image. A more superior-to-inferior trajectory via a posterior transcallosal approach could, in theory, provide better visualization of the inferior portion of the tumor but this brings the deep veins into the surgical corridor. V, top of the cerebellar vermis. **(e)** By retracting the vermis, the right inferior colliculus (*) was seen. The entry into the pons was just inferior to this in the infracollicular entry zone. **(f)** After entry and drainage of the cyst, the left-sided wall of the tumor was clearly seen (middle of the image).

cortex to SSS were encountered posterior to the vein of Trolard. The falx was clearly identified and a retractor blade was placed here to gain slightly more exposure of the left side. The falx was then followed inferiorly towards the tentorium. Once the straight sinus was clearly identified, the tentorial incision was made just lateral to it from approximately the midpoint of the straight sinus all the way to the tentorial incisura. Bipolar cautery was sufficient to stop the venous bleeding from the tentorium found along the incision (**Fig. 25.4b**).

Once the tentorium was cut, attention was focused on the vein of Galen and the origin of the straight sinus. The arachnoid in this area was dissected carefully to expose the vein of Galen with two goals in mind: (1) to see it and then protect it and (2) to visualize the brainstem tectum behind the deep veins (**Fig. 25.4c**). The superior vermis was retracted very gently in an inferior direction, and the right inferior colliculus was clearly visualized (**Fig. 25.4e**).

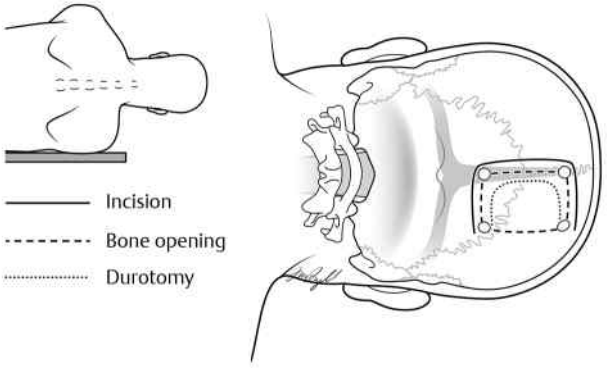
Operative Setup

Positon: lateral with ipsilateral side down

Incision: "U" shape, based on the ipsilateral, down side

Bone Opening: occipital craniotomy with midline burr holes

Durotomy: "U" shape based laterally



At this point, navigation was used to confirm the infracollicular entry zone. Incision near the frenulum veli permitted entry into the top part of the fourth ventricle, and from there, a small incision into the brainstem was made. The trajectory of the dissection was guided at all times by the navigation system and finally, a gush of yellow fluid signaled the location of and entry into the tumor cyst. The endoscope was used to examine the interior of the cyst. On the opposite (left) wall of the cyst, there was a reddish nodule which provided a good starting point for the resection (**Fig. 25.4f**). Back under microscope visualization, the base of this nodule was coagulated with the bipolar, and the nodule was elevated with microsurgical technique. Removal of this nodule led to the deeper solid component of the tumor itself, which was located on the left side of the pons, just inferior to the red nodule. This solid component was carefully dissected away from the brainstem parenchyma, and at times, the endoscopic bipolar was used in a spreading motion to assist this maneuver. There was no discernable "capsule," easily separable from the brainstem, for any portion of the tumor, and dissection proceeded slowly with a "feathered touch" in all quadrants. After the solid mass was removed, the rest of the cyst wall was examined in a 360-degree manor and where it appeared clearly abnormal, was removed as well. At no point did the brainstem neurophysiological signal change during the resection. An edited version of the video from this operation accompanies this text (**Video 25.1**).

The tumor cavity was carefully examined for hemostasis and when necessary, bipolar cautery was used on a very low setting to stop any bleeding. It was unnecessary to close the cut edge of the tentorium. The dura was closed by reapproximating with sutures, and this was augmented with a dural substitute in an onlay manner. The craniotomy bone flap was replaced and the rest of the soft tissue closed. The skin was closed with a running nylon stitch to obtain watertight closure. The postoperative MRI showed a near-total resection with a possible small residual left at the inferior edge (**Fig. 25.5**).

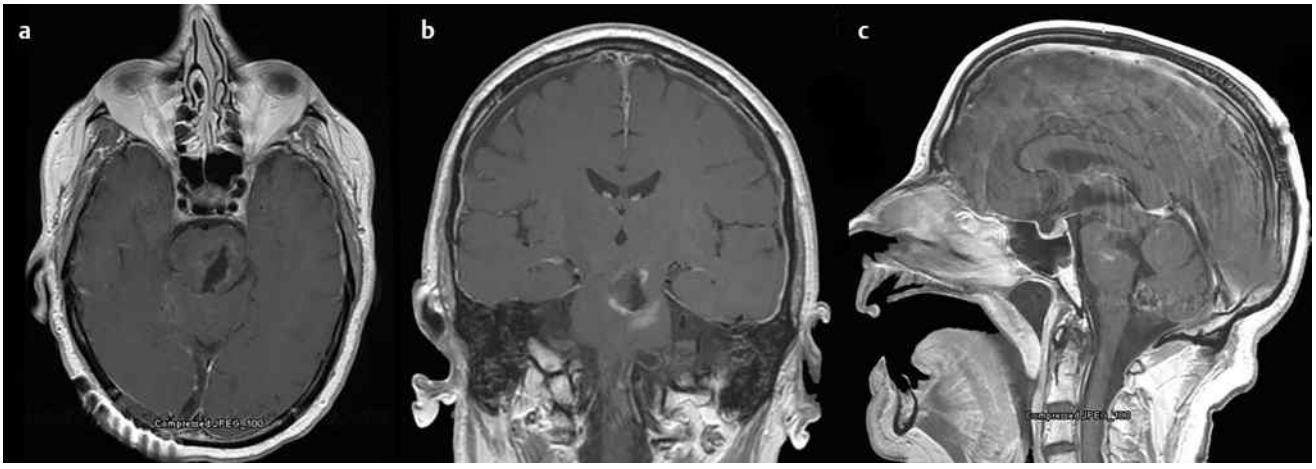


Fig. 25.5 Postoperative MRI. **(a)** Axial, **(b)** coronal, and **(c)** sagittal images showing the right-superior to left-inferior trajectory of the surgical approach. The result was a near-total resection of the tumor, with a thin residual at the inferior extreme. The entry point into the pons is depicted in **(c)** just under the inferior colliculi.

Surgical Pearls

1. As subtleties of the interhemispheric transtentorial approach have already been discussed in a previous chapter, "pearls" here will be limited to this unique case. Keeping the "safe entry zone" at the infracollicular area, a more "top-down" trajectory might have provided better visualization and more exposure to the most inferior aspects of the tumor mass. Such a top-down trajectory could have been provided by a posterior transcallosal approach, but this path would have placed the deep veins and the vein of Galen directly in between the surgeon and the target (**Fig. 25.4**). As such, the more lateral and less "top-down" corridor behind and under the vein of Galen was chosen for working on this tumor. It is critical to have ready an angled endoscope for this operation to overcome any difficulty with seeing the inferior extreme of the tumor.
2. For management of expectations after surgery, the patient's family was counseled beyond the usual extent before this operation. The patient's options were limited at the outset: do nothing and the tumor growth would guarantee certain death; operate and it was highly likely that he would have significant disabilities from surgery. In the end, he chose to do something, rather than to do nothing, in the hope that the "best case scenario" would happen, and he could maintain some functional independence. But whatever the outcome, it would be his family's burden to take care of him, and thus critical that they be engaged in his decision process and aware of its possible outcomes. This is not practicing "defensive medicine," it is simply "good medicine."

■ Aftercare

The patient remained intubated after surgery and had significant quadraparesis, but the left side quickly improved over the first week. His oculomotility was not testable because of impaired arousal at this point. By the second week, it was apparent that he required tracheostomy and gastrostomy placement, but his alertness, attention and left-sided strength continued to improve. By the time he left the hospital between the third and fourth postoperative week, he was answering questions by mouthing words, and regaining movement on the right side.

The histopathological diagnosis was "diffuse midline glioma with histone H3-K27M mutation." This is a tumor entity newly described in the 2016 edition of the World Health Organization (WHO) classification of tumors of the central nervous system, and though tumor morphology is significantly heterogeneous in this classification, tumors in this classification are associated with aggressive clinical behavior.

■ Possible Complications and Associated Management

As ill-defined as the margin of any tumor and the brainstem might be, the radiotherapy that this patient received could only have made it worse. With the myriad of cranial nerve nuclei and fiber tracks densely packed in the pons, the potential complications from this operation are almost innumerable. They certainly include oculomotor palsy, facial paralysis, motor weakness and many more. It was the plan at the outset of the operation to keep the patient intubated after surgery. Predictably, his arousal did not return to normal for quite some time. Performing the tracheostomy and gastrostomy within the week after surgery aided in mobilizing the patient, and prophylaxis against venous thrombosis was important given hemiparesis that persisted.

Perspective

Robert F. Spetzler

■ Introduction

Although the approach presented in the previous section is quite reasonable, I would not have chosen this route. However, before discussing the surgical approach, the decision to proceed with surgery deserves a few comments.

The discussion of the differential diagnosis of this lesion is most appropriate. The likelihood that a diagnosis can be obtained from the cyst fluid is remote, and therefore the risk of proceeding with stereotactic aspiration would not have been an option I would have presented to the patient. Additionally, at the initial presentation, I would have considered two options, open surgery or focused radiation. Because the diagnosis of hemangioblastoma was remote, and a diagnosis of either metastatic or aggressive primary tumor was the most likely, I would have suggested Gamma Knife (Elekta AB, Stockholm, Sweden) radiosurgery as the best treatment course with the least risk and greatest potential benefit to the patient.

With recurrence and significant deterioration 1 year later, the patient was confronted with a difficult choice of a risky surgery or further palliative treatment, with the latter being unlikely to produce any benefit. The will to live is inherently strong in our species and most patients will grasp at any straw offering a chance of meaningful survival, even when the odds are slim. The risks and benefits were appropriately presented, and the patient chose to proceed with surgery. The surgery was elegantly performed, and the goals of a definitive diagnosis and maximal resection were achieved. Nonetheless, the patient paid a significant price in the form of a postoperative neurologic deficit, and the question of a quality life-extension benefit remained unanswered.

Although I, too, would have suggested surgery for this patient, my approach would have been a lateral supracerebellar infratentorial (SCIT) route. The advantages of this approach include no need for retraction, thus minimizing the risk of homonymous hemianopia, no risk of injury to the critical veins, and a lateral entry point into the lesion that is extremely well tolerated and that avoids the quadrigeminal plate. The following case illustrates this approach and the advantages inherent to it.

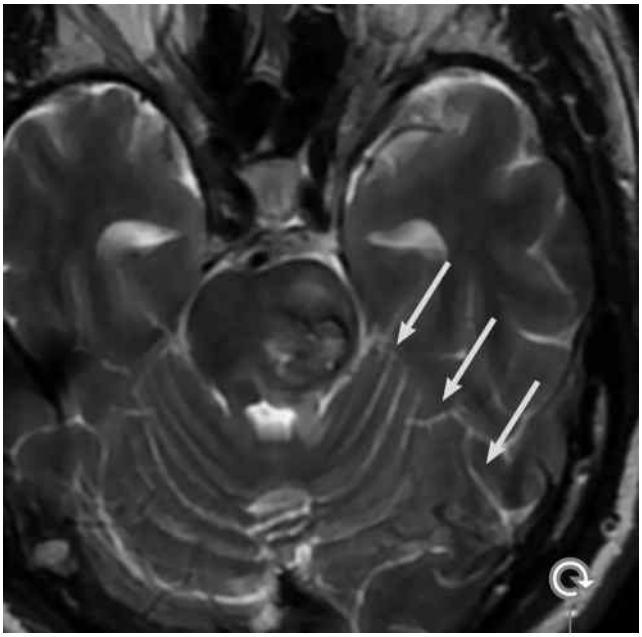


Fig. 25.6 Axial magnetic resonance image demonstrating a large pontine cavernous malformation with surrounding edema. Arrows depict the location of the tentorium separating the supratentorial and infratentorial compartments. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

■ Case Presentation

A 62-year-old man presented with acute onset of headache, right-sided weakness, diplopia, and dysmetria. MRI revealed a hemorrhagic lesion in the pons extending into the midbrain, and the diagnosis of a cavernous malformation was easily established (**Fig. 25.6, Fig. 25.7**). Surgery was uneventful (**Figs. 25.8**–**25.13**), and the patient made an excellent recovery (**Fig. 25.14**).

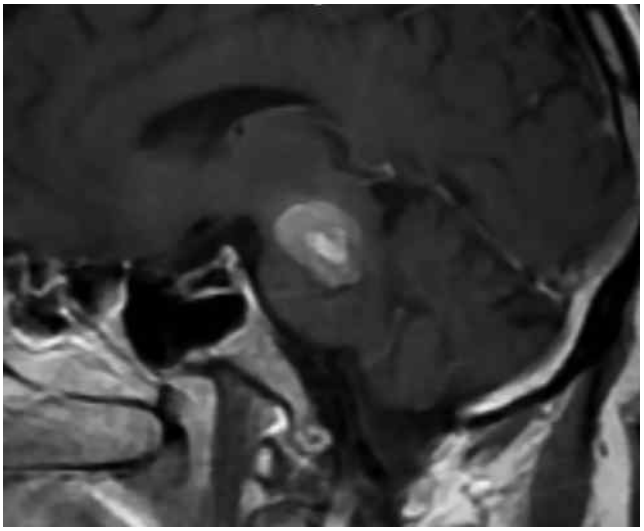


Fig. 25.7 Sagittal magnetic resonance image demonstrating extent of cavernous malformation and hemorrhage. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

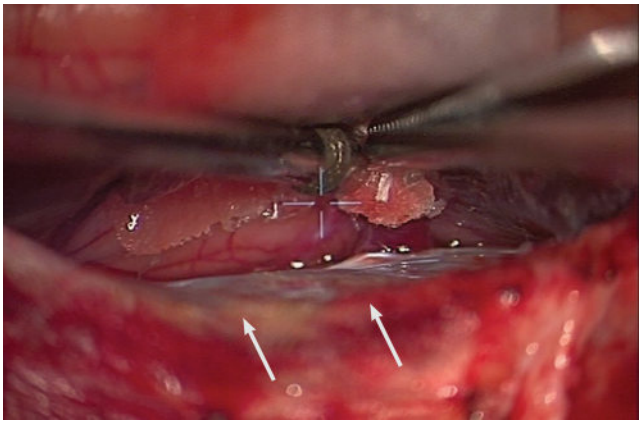


Fig. 25.8 With the patient in the supine position and the head turned, the cerebellum is gently elevated from the tentorium and the lateral sinus to obtain access to the ambient cistern. No retractors are required for the transverse sinus (*arrows*), which is retracted naturally by gravity due to the patient's position. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

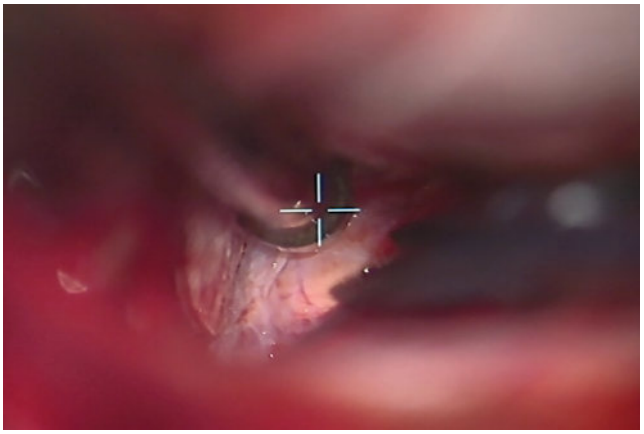


Fig. 25.9 The arachnoid membrane covering the ambient cistern is opened. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

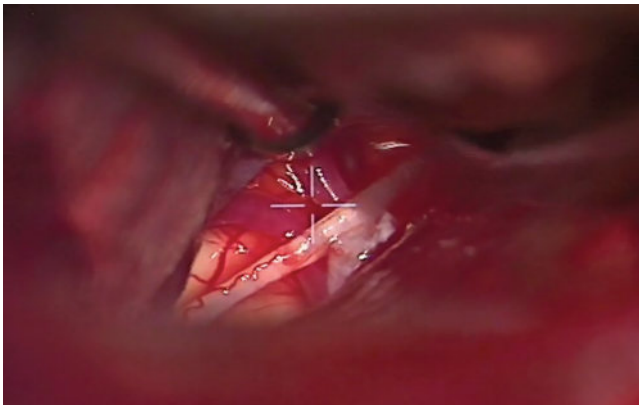


Fig. 25.10 After the arachnoid membrane is opened, the trochlear nerve (cranial nerve IV) and the branches of the superior cerebellar artery become visible. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

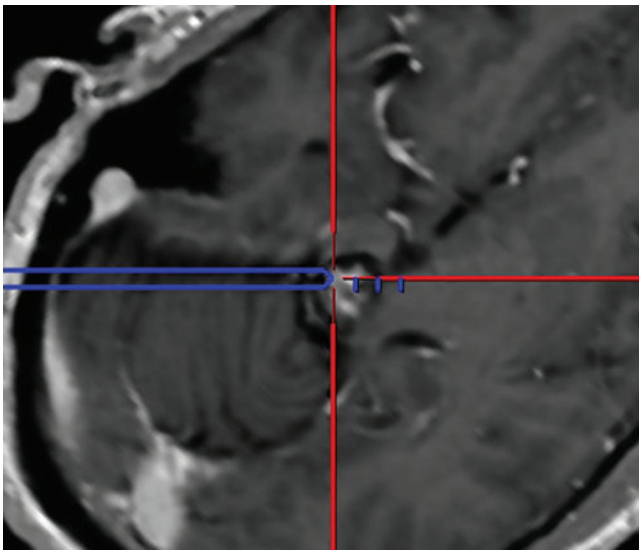


Fig. 25.11 Essential image guidance demonstrates the trajectory as well as the lateral approach. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

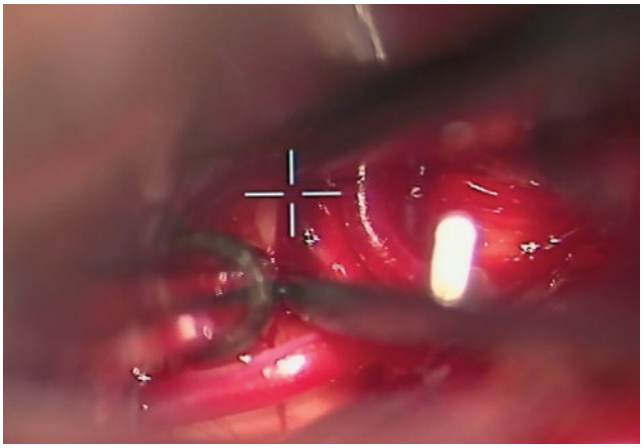


Fig. 25.12 The midbrain is opened laterally through the medial lemniscus. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

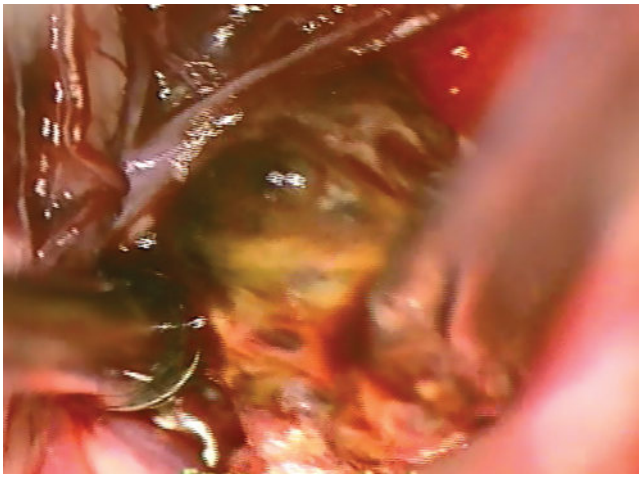


Fig. 25.13 The cavernous malformation is mobilized and removed. No rigid retraction was used at any point in the surgery. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

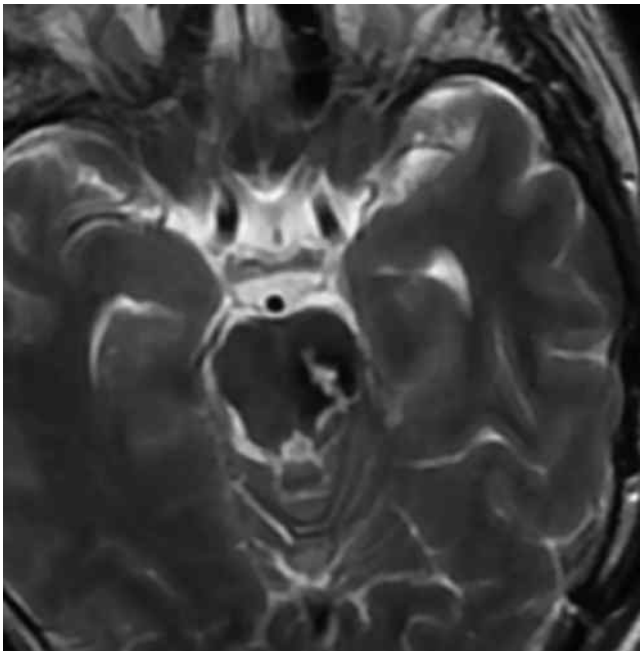


Fig. 25.14 Postoperative magnetic resonance image demonstrates the entry point to the cavernous malformation and documents complete resection of the lesion. (Used with permission from Barrow Neurological Institute, Phoenix, AZ.)

■ Commentary

The suitable approaches to lesions in this location have been fully discussed in the previous section, and the advantages of using the SCIT are multiple: (1) supine (or lateral, depending on neck flexibility) patient positioning avoids venous congestion; (2) comfortable sitting position for the surgeon; (3) straightforward route to the lesion along the tentorium; (4) no risk to the deep venous drainage; (5) no risk to the occipital lobe; (6) great tolerance with the lateral midbrain opening to avoid neurologic deficit; and (7) ability to extend superiorly to the level of the foramen of Monro by cutting the tentorium and extending the incision far inferiorly tothe pons.

Important pearls include skeletonizing the transverse sinus so that it can be retracted (**Fig. 25.8**) to avoid any overhang, resulting in better visualization and less need for deep retraction. Additionally, lighted instruments are helpful in allowing maximal visualization in small, deep, recessed corridors.

■ Recommended Reading

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26 Brainstem (Pontomedullary)

Karolyn Au and Jacques J. Morcos

Keywords: cavernous malformation, far-lateral approach, occipital condyle, two-point method, endoscopic endonasal transclival approach

■ Case Presentation

A 31-year-old previously healthy woman experienced sudden onset of headache, as well as right hand and arm tingling. One day later, she developed slurred speech and right-sided clumsiness. These symptoms progressed over two days, and she presented to the emergency department. Physical examination revealed normal sensation testing, symmetric facial movement, and midline uvula and tongue protrusion. However, she was hemiparetic on the right with 4/5 power in the upper extremity and 4+/5 power in the lower extremity. A noncontrast computed tomography (CT) scan of the head was performed, demonstrating an area of hyperdensity in the ventral pons, eccentric to the left (**Fig. 26.1**).

Questions

1. Based on the CT images, what additional investigations should be obtained?
2. What anatomic structures are responsible for the patient's neurological findings?
3. Do you think this patient required surgical intervention? Or, is observation a better option for her?

Fig. 26.1 Preoperative CT. Axial noncontrast CT demonstrated an area of hyperdensity within the anterior pons.

■ Diagnosis and Assessment

The differential diagnosis for the lesion seen on CT includes cavernous malformation (also known as a "cavernoma"), arteriovenous malformation, thrombosed vertebrobasilar aneurysm with brainstem compression, and hemorrhagic intra-axial tumor such as metastasis or high-grade glioma. Further investigations should therefore include contrast-enhanced MRI as well as angiography.

MRI of the brain with gadolinium contrast demonstrated a nonenhancing intra-axial lesion with mixed densities on both T1- and T2-weighted sequences, consistent with a cavernous malformation (**Fig. 26.2**). MR angiography obtained at the same time did not show any filling of the lesion, in keeping with the diagnosis of cavernoma. A digital subtraction angiogram performed to definitively rule out a high-flow vascular lesion did not demonstrate any abnormality. An MRI of the body was obtained to look for additional vascular or neoplastic lesions, as would be possible in the setting of cavernous malformation or metastasis. This was also unremarkable.

Understanding the natural history of cerebral cavernous malformations has been challenging due to heterogeneity in populations, diagnostic criteria, methods of assessment, and outcome definitions among different studies. Furthermore, most available series are retrospective in nature, making the true risk of hemorrhage difficult to describe. Nevertheless, some patterns are evident: the presence of prior hemorrhage has consistently been shown to be a significant risk factor for future symptomatic hemorrhage, and numerous series have demonstrated the brainstem location to be associated with higher risk for hemorrhage. A meta-analysis of individual patient data from existing series found that, compared to malformations in non-brainstem locations that presented without hemorrhage or focal neurologic deficit (FND), lesions in the brainstem had a hazard ratio of 1.8. Moreover, cavernous malformations in the brainstem that had previously presented with hemorrhage or FND had a hazard ratio of 10.2 for hemorrhage at 5 years. Looking at the data another way, the 5-year risk of hemorrhage in this group was a remarkable 30%.

This patient had presented with a solitary cavernous malformation in the lower pons with acute symptomatic hemorrhage. With time, the mass effect and edema resulting from acute hemorrhage were expected to decrease; indeed, the patient made a near-complete recovery over the following month, demonstrating only trace residual weakness in the right arm. With the natural history in mind, we recommended surgical intervention to her. The goal of surgery was to remove the malformation to prevent repeat hemorrhage with neurologic deterioration, while avoiding new neurologic deficits.

■ Anatomical and Therapeutic Considerations

The malformation occupied the caudal portion of the basis pontis, from mid-pons rostrally, to below the pontomedullary

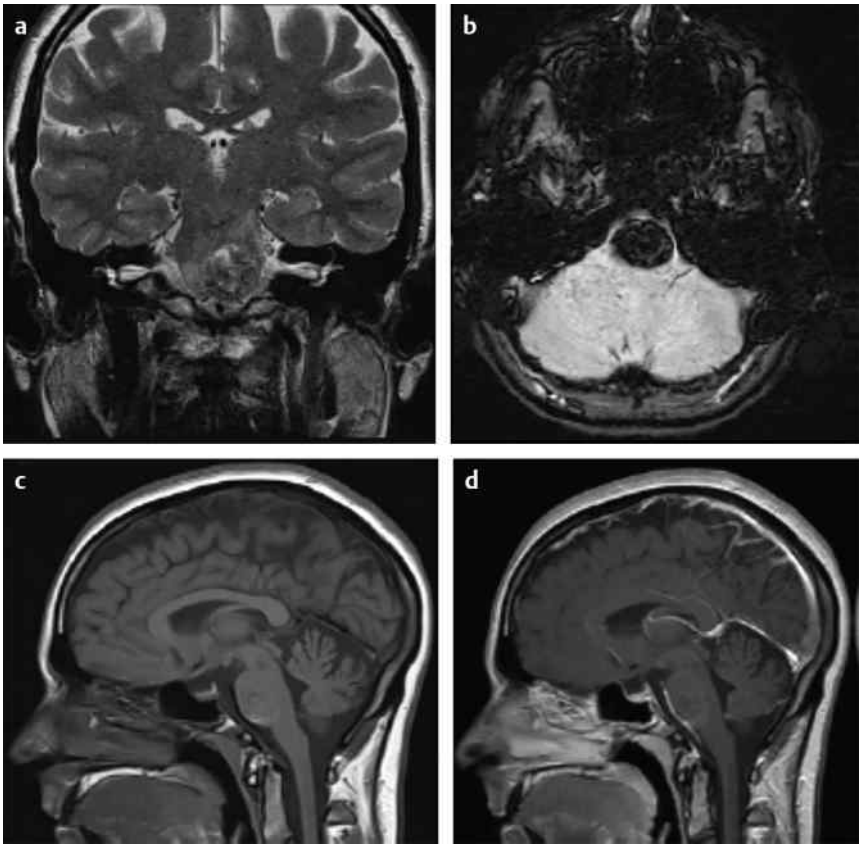


Fig. 26.2 Preoperative MRI. (a) Coronal T2-weighted sequence showed the caudal extension of the lesion below the pontomedullary junction. **(b)** Blooming artifact on susceptibility-weighted imaging consistent with acute hemorrhage is seen on the CT. **(c)** T1-weighted precontrast and **(d)** postcontrast sequences demonstrated no gadolinium contrast enhancement within the lesion.

junction caudally. Bundles of corticospinal tract fibers occupy much of the pontine tegmentum, surrounded by transverse pontine fibers, while pontine nuclei reside deep to these white matter tracts (**Fig. 26.3**). The malformation was eccentric to the left, explaining the patients' right-sided hemiparesis. It extended to the pial surface at the ventrolateral aspect of the pons, making this the safest point of entry to avoid traversing normal brainstem parenchyma. The trigeminal nerve roots approach the mid-pons from a paramedian ventral aspect and enter the brainstem at approximately the level of the rostral pole of the malformation. The facial and vestibulocochlear nerves exit at the pontomedullary junction and course anterolaterally through the internal auditory canal; the malformation extended caudally well beyond this level.

The trunk of the basilar artery ascends over the ventral surface of the pons, to which it supplies perforating arteries. A prominent venous structure was seen at the ventral aspect of the malformation, adjacent to the basilar artery.

Options for Approach

Options to access the ventral pons include an anterior petrosal approach and a posterolateral approach. The first approach requires a temporal craniotomy and anterior petrosectomy. Extradural elevation of the temporal lobe is performed with care to avoid excessive traction on the greater superficial petrosal nerve and the vein of Labbé. This technique provides direct access to the ventrolateral brainstem surface, allowing the surgeon to work medial to the facial/vestibulocochlear nerve complex. The disadvantage of this approach, however, is its limited reach for lesions extending below the level of the internal auditory canal. Alternatively, a posterior petrosectomy, by drilling out the retrolabyrinthine portion of the petrous temporal bone, would certainly expose our entire target area. However, this extensive removal of bone is time consuming and unnecessary.

Approach of Choice

Although a traditional retrosigmoid approach would be too limited and provide insufficient anterior trajectory to reach our target, this opening can be modified to include drilling of the foramen magnum rim up to the occipital condyle. Surgical access through the resultant far-lateral opening would provide the surgeon with good access through the posterolateral corridor to the entire lower brainstem without massive bone removal.

Questions

1. Which approach would you choose for the patient?
2. What anatomic variants of bone and vasculature at the craniocervical junction can be encountered while performing a far-lateral exposure?
3. Some cavernous malformations are associated with a developmental venous anomaly. How are these venous structures managed during resection of the malformation?

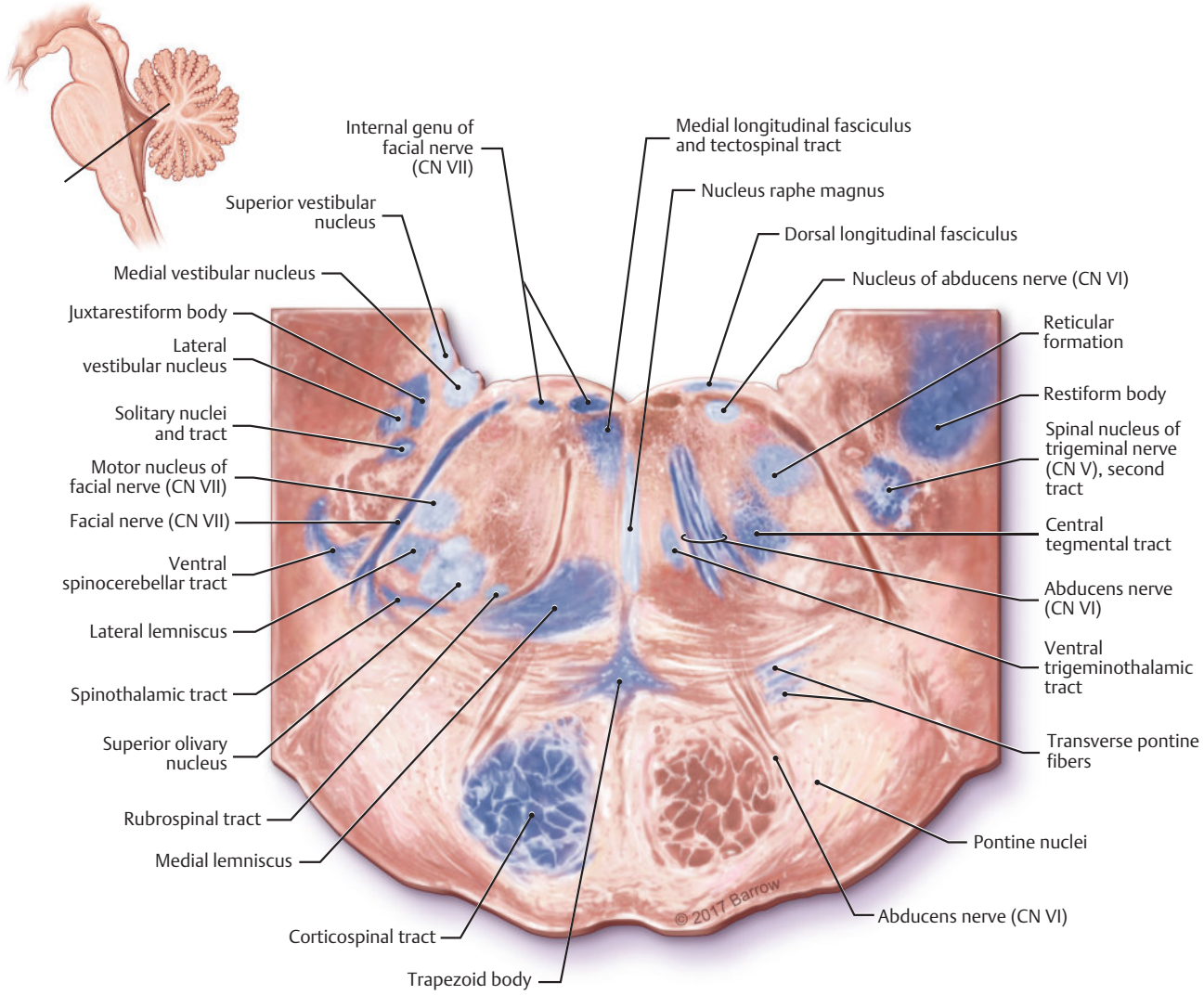


Fig. 26.3 Transverse section of the caudal pons through the facial colliculus.

■ Description of the Technique

A far-lateral approach was chosen for this patient, modified to include a suboccipital/retrosigmoid craniotomy and opening of foramen magnum to the occipital condyle (see **The Three Approach Elements**). A C1 laminectomy was not needed. Standard anesthetic techniques for brain relaxation were essential, and electrophysiologic monitoring of somatosensory, motor, and brainstem-auditory evoked potentials was used to provide early warning of neurologic compromise.

The Three Approach Elements

Corridor: posterior (inferior) lateral

Craniotomy: retrosigmoid

Modifiers: minimal condylectomy, expose sigmoid

To avoid the risk of venous air embolism associated with the sitting position, the patient was placed on her right side (i.e., left-side up) in the three-quarter-prone position. This position also allowed gravity to draw the cerebellar hemisphere away from the surgical corridor. The chest was supported with gel rolls below the axilla, and the right arm was held in a sling suspended from the end of the operating table. A Mayfield clamp was applied and four cardinal movements were used to achieve ideal positioning of the head: (1) anterior flexion to expose the suboccipital region, (2) right lateral flexion to create working room beside the left shoulder, (3) rotation of the face toward the floor to elevate the left suboccipital surface, and (4) upward translation to open the left atlanto-occipital joint. Without applying excessive force on the brachial plexus, the left shoulder was gently pulled downward and forward. The patient was secured to the table to allow for side-to-side rotation, and the head elevated by placing the table in reverse Trendelenberg. The

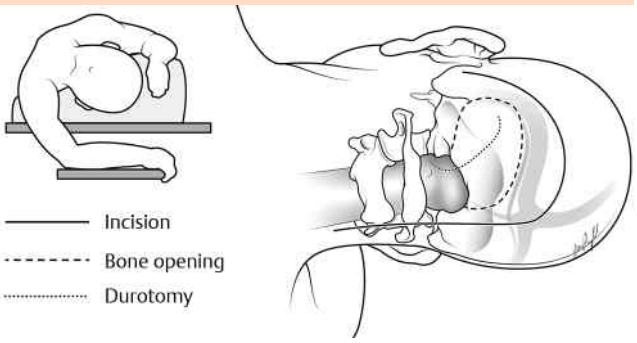
Operative Setup

Position: three-quarter prone, right-side down for left-sided approach

Incision: "hockey stick" midline C2 to above inion, laterally to mastoid, inferiorly to mastoid tip

Bone opening: suboccipital craniotomy, laterally to sigmoid, caudally to condyle

Durotomy: wide "U" shape based laterally



patient was registered to preoperative imaging on the frameless stereotactic guidance system (see **Operative Setup**).

The incision began at the level of the C2 spinous process, extended in the midline to 2 cm above the inion, continued laterally to the level of the mastoid process, and finally turned inferiorly to end at the mastoid tip. Dissection in the midline reduced muscle trauma, and with preservation of a cuff at the superior nuchal line for closure, the muscle was elevated laterally to identify the mastoid process and digastric groove and caudally to expose the foramen magnum rim. The posterior arch of C1 was exposed in the midline, allowing for identification of bony landmarks before the dissection was continued laterally to uncover the vertebral artery. Attachments to the C2 spinous process did not need to be disrupted. The muscle flap was firmly retracted inferolaterally with large hooks to minimize its bulk.

The initial drilling began within the angle of the transverse–sigmoid junction, aiming toward the digastric groove to create a trough. Using a large diamond burr on a high-speed drill, the dura at the medial edge of the sigmoid sinus was exposed. After dissection of the epidural space, a craniotomy flap was elevated, maximizing exposure of dura caudally. Further drilling was carried out to expose the entire medial edge of the sigmoid sinus, and to open the foramen magnum from the right side of midline to the edge of the left occipital condyle. As drilling proceeded toward the condyle, the vertebral artery was protected using gentle retraction by the surgical assistant. Bleeding from the posterior condylar emissary vein was controlled with tamponade and hemostatic agents, and signified the lateral limit of the exposure. In this patient the needed rostral trajectory was achieved without removal of the C1 lamina.

A small opening was initially made in the dura below foramen magnum, allowing cerebrospinal fluid (CSF) egress to relax the posterior fossa. The dural opening was then continued rostrally over the left cerebellar hemisphere, with a short transverse limb at each end to form a wide "U" shape based laterally. Stay sutures were placed at the base of the dural flap to ensure maximal lateral retraction. The rootlets of glossopharyngeal and vagus nerves were identified, and the arachnoid around them sharply dissected to allow the cerebellum to fall away. The dissection continued rostrally toward the facial/vestibulocochlear nerves and the trigeminal nerve.

The neuronavigation probe was placed on the ventrolateral pons above and below the VII/VIII nerve complex, confirming that the malformation was centered at the more caudal level. The pontine surface was inspected in the window between the VII/VIII and IX/X nerves, and a large vein and the abducens nerve were identified. A region lateral to abducens nerve and caudal to the vein with few pial vessels was cauterized and opened sharply in parallel to the transverse pontine fibers (**Fig. 26.4**, **Fig. 26.5a**). The hematoma cavity was entered and separation of the malformation from surrounding gliotic brain parenchyma was carried out using a combination of sharp and blunt dissection (**Fig. 26.5b**). Gripped with forceps, the malformation was pulled into the cavity, with countertraction applied to the adjacent brain (**Fig. 26.5c**). Due to the location, the lesion was removed piecemeal. The cavity was inspected to confirm that no malformation remained (**Fig. 26.5d**). **Video 26.1** demonstrates the technique used in this operation.

The dura was closed primarily with suture and reinforced with onlay of synthetic graft material. Exposed air cells at the craniotomy edge were thoroughly waxed to avoid CSF leak, and the occipital craniotomy flap was replaced. The muscle was closed in multiple layers, facilitated by the cuff preserved on the occipital bone. Running nylon suture was used to close the skin in watertight fashion. Postoperative MRI showed complete removal of the malformation (**Fig. 26.6**).

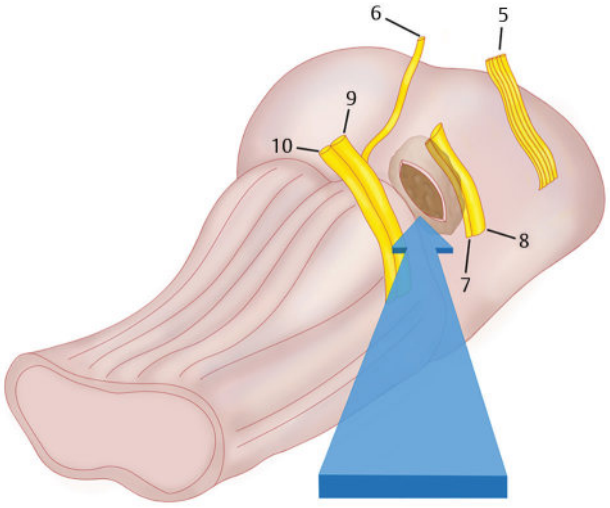


Fig. 26.4 Illustration of the brainstem entry site. The malformation occupied the caudal portion of the basis pontis, from mid-pons to below the pontomedullary junction. It extended to the pial surface at the ventrolateral aspect of the pons, making this the safest point of entry. An area lateral to abducens nerve and just medial to the VII/VIII nerve complex was opened sharply in parallel to the transverse pontine fibers. 5, trigeminal nerve; 6, abducens nerve; 7, facial nerve; 8, vestibulocochlear nerve; 9, glossopharyngeal nerve; 10, vagus nerve.

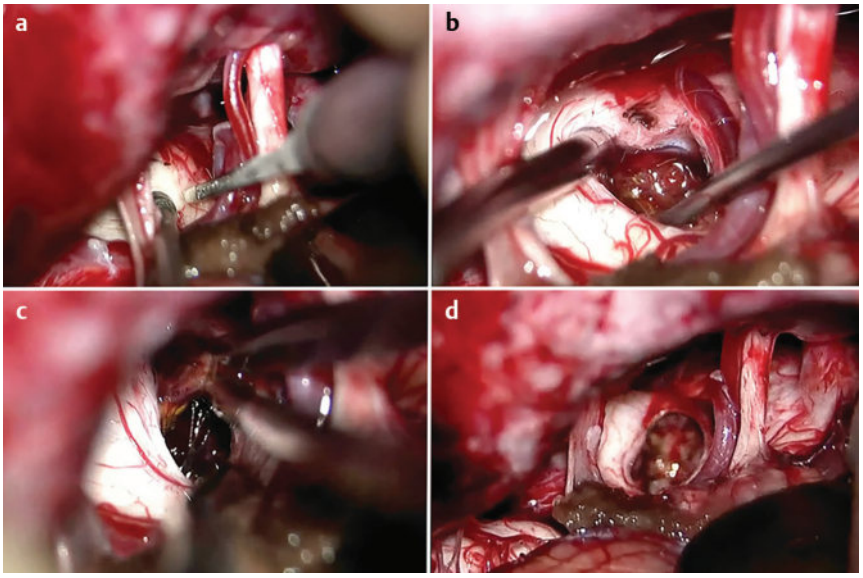


Fig. 26.5 Intraoperative images. (a) A wide view of the lateral brainstem has been provided by the far-lateral craniotomy, allowing a trajectory to the lower pons from below and medial to VII–VIII complex. A transverse incision was made in the lateral pons where the malformation most closely approaches the pia, parallel to the transverse pontine fibers. **(b)** The microdissector was used to sweep around the edge of the malformation, separating it from the brainstem parenchyma. **(c)** The malformation was grasped and mobilized into the resection cavity, while the suction applies countertraction. **(d)** The resection cavity was seen below cranial nerves VII to VIII complex.

Fig. 26.6 Postoperative MRI. Axial T2-weighted MRI obtained on the second postoperative day showed complete resection of the malformation.

Surgical Pearls

1. Anatomic variations such as an incomplete lamina of C1 or C1 assimilation should be identified on the preoperative imaging. The vertebral artery may take a tortuous course above C1 before it penetrates the dura, or be constrained by an ossified atlanto-occipital membrane. Infrequently, the posterior inferior cerebellar artery or the posterior spinal artery may arise from the extradural portion of the vertebral artery, and must not be mistaken for muscular branches and divided. These variations must be identified preoperatively to avoid vascular injury.
2. The goal of the far-lateral approach is to create an inferolateral corridor to the anterior brainstem while avoiding retraction. At every step, key details facilitate this goal: the use of large hooks for muscle retraction, instead of self-retaining retractors, decreases the bulk that can impinge on anterior access; bone at the foramen magnum must be drilled to the occipital condyle, ensuring that a small lip does not remain to protrude into the exposure. The dura should be retracted with sutures placed at its lateral base to fully utilize the bone opening.
3. As there is little room for dissection around the periphery of a lesion within the brainstem, a cavernous malformation may need to be removed piecemeal. Evacuation of the hematoma creates room to manipulate the malformation, which can be pulled into the cavity while the brain parenchyma is dissected away. A combination of sharp and blunt dissection is used, depending on the interface between the malformation and the surrounding brain. Associated developmental venous anomalies drain normal brain and must be preserved to avoid venous congestion and infarction.

■ Aftercare

Immediately following surgery, the patient was neurologically unchanged compared to before surgery, with subtle weakness of the right arm. She was independently ambulatory and was discharged for home 3 days later. The pathology demonstrated thin-walled, dilated vessels surrounded by hemosiderin and gliosis, consistent with cavernous malformation. Follow-up imaging 1 year later demonstrated no sign of residual (**Fig. 26.7**).

■ Possible Complications and Associated Management

There is great debate in the literature about how much of the occipital condyle can be removed—if any—without destabilization of the atlanto-occipital joint. The drilling of the occipital

Fig. 26.7 Follow-up MRI. Axial T2-weighted MRI obtained at 1-year follow-up demonstrated no residual malformation.

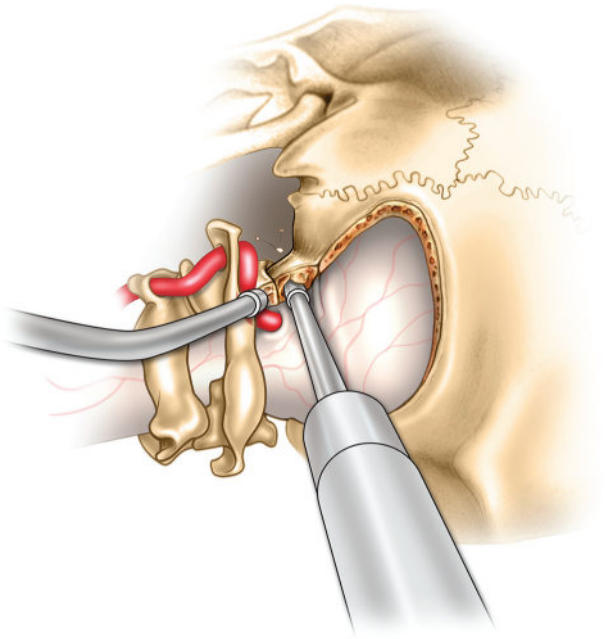


Fig. 26.8 The far-lateral retrocondylar and transcondylar approaches. The amount of the condyle removed must be tailored to each individual patient's lesion, its size, and location.

condyle must therefore be tailored case-by-case, and must cease when sufficient exposure is obtained to any specific lesion. For this patient, only a small amount of the condyle needed to be removed to reach the cavernoma at the lateral pontomedullary junction. The drilling stopped when the condylar emissary vein was reached and only a small fraction of the condyle was removed. Although conflicting data exists regarding maximal condylar removal, most investigators would agree that less than one-thirds is safe with respect to joint stability. For targets that require a more lateral-to-medial trajectory, when more of the occipital condyle needs to be removed, careful postoperative evaluation of the atlanto-occipital joint would be necessary to determine the need for instrumented fusion and stabilization.

Retrocondylar versus Transcondylar versus "Enough Condylar"

When it comes to working around the occipital condyle, how much needs to be removed (**Fig. 26.8**)? To answer this question more in depth, a couple more relevant facts need to be mentioned. Extra-axial tumors in and around the brainstem expand space and therefore generate a path to their own resection. On the other hand, intra-axial tumors and vascular lesions, such as aneurysms and arteriovenous malformations, leave the surrounding cisternal anatomy, by and large, undisturbed, and consequently the cone of surgical viewing remains narrow. It is because of this simple contrast that a counterintuitive consequence arises: generally, less bone needs to be drilled for a meningioma than an aneurysm of the same location. In other words, the more shift the lesion has caused, the less space the surgeon has to create for exposure. A related concept is the well-recognized trend that accumulated surgical experience lessens the required amount of exposure. A more experienced surgeon will acquire the skills of "efficient surgical space management" and become comfortable with incorporating a "transtumoral" approach in his/her armamentarium. Unless the pathology is within the condyle, such as occurs in chordomas or chondrosarcomas, it is highly unlikely that we would drill significantly into the condyle for any/most lesions of the brainstem or craniovertebral junction. The workhorse approach to this region remains the juxta (retro)condylar approach, and not the transcondylar approach.

Perspective

Frederick L. Hitti, Omar Choudhri, and John Y.K. Lee

■ Introduction

Cavernous malformations of the brain are rare lesions with a prevalence of less than 1%. However, when they exist and hemorrhage in the brainstem, they can produce devastating neurological injury. Furthermore, as the authors of the previous section have pointed out, once a cavernoma has bled, it becomes more likely to bleed again, and rebleed rates can be as high as 35% per year. Taken together, this makes brainstem cavernomas particularly challenging lesions for modern neurosurgery. They are difficult to access, often requiring working through deep corridors. Damage to the surrounding normal brain, whether through repeated hemorrhage or surgical manipulation, can result in profound neurological injury.

The resection of a pontomedullary cavernoma using microsurgical techniques was discussed earlier. Since the malformation came to the surface at the ventrolateral surface of the brainstem, a posterolateral corridor was used for its removal. Had the lesion been positioned slightly more anteriorly, and surfaced the brainstem purely on the ventral aspect, this trajectory would have been very difficult. In the past decade, endoscopic resection of deep intracranial lesions including cavernomas has been gaining acceptance, and the endoscopic transclival technique may be particularly useful for cavernomas positioned ventrally in the brainstem.

In this section, we present the endoscopic technique and discuss the advantages and disadvantages of this approach.

■ Case Presentation

A 60-year-old woman presented to our clinic with right hemiparesis. She had a medical history notable for a brainstem glioma located in the medulla that had been diagnosed 11 years prior to the current presentation. She was initially treated with radiation. Three years later, when she experienced neurological decline, she was advised to receive further treatment with chemotherapy. However, she continued to decline in a stepwise fashion. Brain MRI was obtained that revealed a nonenhancing, noninfiltrative lesion in the ventral medulla. Furthermore, gradient echo sequence revealed hemorrhage within the lesion (**Fig. 26.9**). These imaging findings were consistent with diagnosis of a radiation-induced cavernoma. Additionally, the patient's clinical history of a stepwise decline supported the diagnosis of a cavernoma with multiple hemorrhagic events.

The patient presented to us following an abrupt neurological decline resulting in a dense right hemiparesis. The patient, at this point, was unable to ambulate independently and required a wheelchair. On neurological examination, the patient was alert and oriented with no cranial nerve deficits. Her right upper extremity was noted to have 2/5 strength proximally and 3/5 strength distally. She had 2/5 strength throughout her right lower extremity, and she had 5/5 strength in her left upper and lower extremities.

■ Anatomical and Therapeutic Considerations

As detailed above, the hemorrhagic lesion was located in the ventral medulla. On the brain MRI, the lesion measured 8 × 9 × 10 mm and approached the surface of the medulla ventrally near the cervicomedullary junction. Given the patient's profound decline and relatively high risk of rehemorrhage, we offered surgical resection of the lesion.

The "two-point" method for determining surgical approach to brainstem lesions has been promulgated by Dr. Brown and his colleagues. While some surgeons can intuit this approach, formalization of the concept has facilitated surgical planning for these lesions. To utilize this method, one point is placed at the center of the lesion and another point is placed where the lesion presents to the surface of the brainstem. A line is then drawn between these two points and extended outward toward the cranial surface. The intersection of this line with the skull dictates the surgical approach. The goal of this method is to minimize brain retraction and minimize/eliminate destruction of normal brain tissue. By placing a point where the lesion approaches the surface of the brainstem, the surgical resection can begin at this point and removal/disruption of normal brain is avoided. The goal of placing the second point at the center of

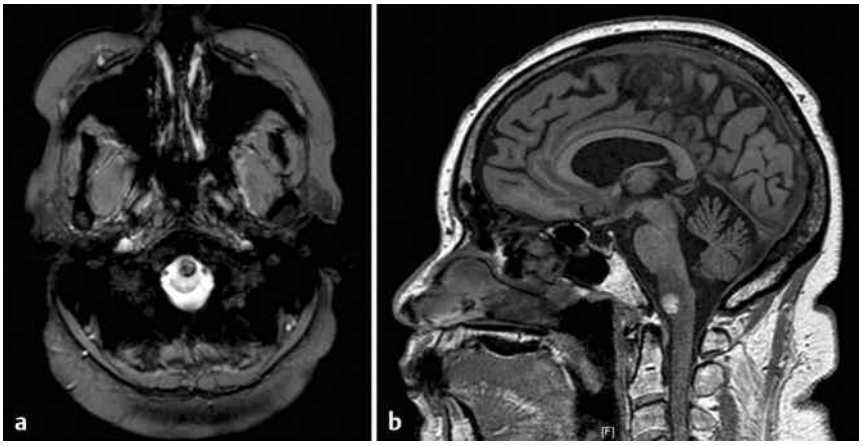


Fig. 26.9 Preoperative brain MRI. (a) Axial gradient echo image demonstrating a hemorrhagic lesion in the medulla. **(b)** Sagittal T1 image (*without* gadolinium) revealing a well-circumscribed lesion in the ventral medulla. (Reproduced with permission from Nayak NR, Thawani JP, Sanborn MR, Storm PB, Lee JY. Endoscopic approaches to brainstem cavernous malformations: Case series and review of the literature. Surg Neurol Int. 2015; 6:68.)

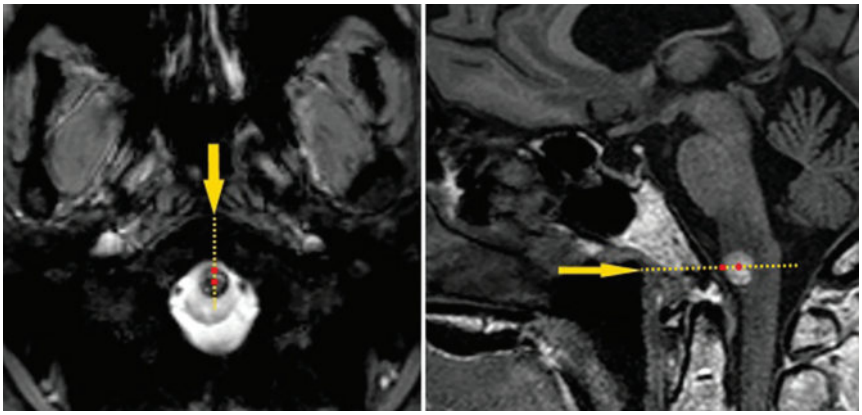


Fig. 26.10 The two-point method. Close-up of **Fig. 26.9** with the *red points* indicate the center of the lesion and where the lesion presents to the surface. A *yellow dashed line* is drawn between these points to reveal the optimal surgical approach, marked by a *yellow arrow*. (Reproduced with permission from Nayak NR, Thawani JP, Sanborn MR, Storm PB, Lee JY. Endoscopic approaches to brainstem cavernous malformations: Case series and review of the literature. Surg Neurol Int. 2015; 6:68.)

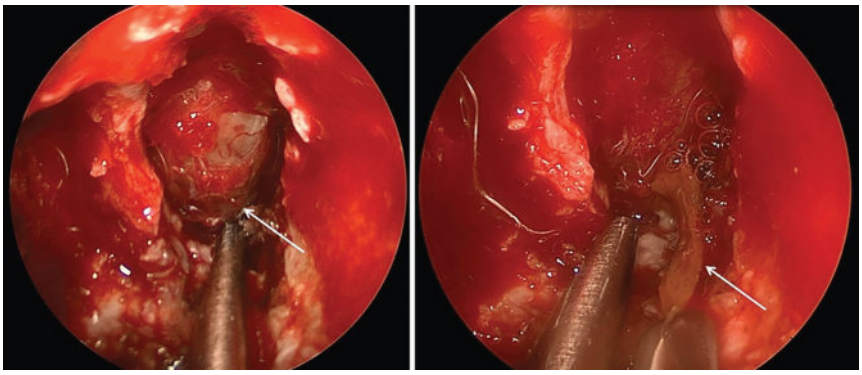


Fig. 26.11 Intraoperative endoscopic images. View of the ventral medulla. Area of hemosiderin stained pia is denoted with an arrow (*left*). After the lesion was debulked, the capsule, indicated with an arrow, was removed (*right*). (Reproduced with permission from Nayak NR, Thawani JP, Sanborn MR, Storm PB, Lee JY. Endoscopic approaches to brainstem cavernous malformations: Case series and review of the literature. Surg Neurol Int. 2015; 6:68.)

the lesion is to minimize brain retraction. If the center of the lesion is approached directly, the lesion may be resected with minimal retraction.

Applying the two-point method on our patient's brainstem lesion, we opted for an endoscopic endonasal transclival approach (**Fig. 26.10**). This method would allow us to approach the lesion ventrally, at the point at which the lesion presented to the surface of the brainstem. The trajectory also allowed us to resect the lesion with no brain retraction, because the center of the lesion was in line with the entry point.

■ Description of the Technique

For endonasal cases at our institution, we work jointly with our otolaryngology colleagues (ear, nose, and throat [ENT]). The ENT surgeon performs the initial approach, and once access to the sphenoid sinus is obtained, they were joined by the neurosurgical team. The patient was placed supine with her head on a donut and a frameless navigation system was registered. The ENT team raised nasoseptal flaps in preparation for the closure. The left middle turbinate and the posterior nasal septum were removed. A high-speed drill was then used to gain access to the sphenoid sinus. Ethmoidectomies were also performed for better exposure. Throughout this process, the surgical navigation system was used to confirm anatomical landmarks.

At this point, the ENT surgeon moved to the patient's left side, while the neurosurgeon operated from the patient's right side. The ENT surgeon operated the endoscope through the left nostril to provide visualization while the neurosurgeon used a high-speed drill and suction to proceed with drilling of the clivus. Both cutting and diamond drill bits were employed for this process. Bone was removed in the caudal direction until the ring of C1 was visualized. The surgical navigation system and an intraoperative lateral X-ray were used to confirm the location of this anatomical landmark. The floor of the sella was the superior limit of our exposure while the lateral limits of the exposure were delineated by the internal carotid arteries and occipital condyles. The apical ligament was transected so that the exposure could proceed down to the foramen magnum.

The dura was opened with a midline incision caudal to the basilar artery to expose the medulla. An area of hemosiderin-stained pia was noted and determined to be the ventral presentation of the lesion (**Fig. 26.11**). Once the pia was opened, necrotic/hemorrhagic tissue was expressed from the center of the lesion. With the core of the lesion debulked, we were able to identify a capsule. Yaşargil forceps were used to remove the capsule en bloc (**Fig. 26.11**). The resulting cavity did not appear to contain any residual abnormal tissue.

Following resection of the lesion, we proceed with closure. Meticulous closure is required for these cases, as CSF leaks are not uncommon. Fat and fascia lata were harvested from the patient's right thigh and were used to repair the defect. The vascularized nasoseptal flaps raised at the start of the case were then placed over the fat and fascia lata grafts to complete the closure.

■ Aftercare

We obtained a postoperative brain MRI to determine the extent of resection (**Fig. 26.12**). With our endoscopic approach, we were able to achieve gross total resection of the lesion. The patient remained at her neurological baseline postoperatively, and at her 3-month follow-up, there were no surgical complications.

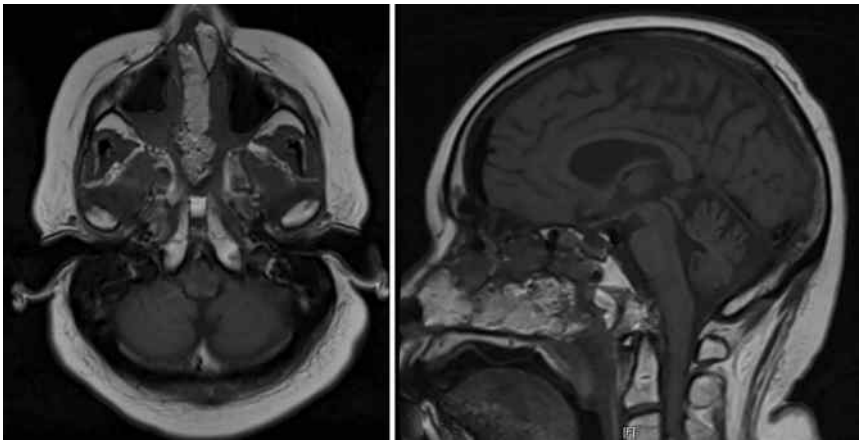


Fig. 26.12 Postoperative brain MRI. Axial (*left*) and sagittal T1 sequences (*right*), demonstrating gross total resection of the lesion. (Reproduced with permission from Nayak NR, Thawani JP, Sanborn MR, Storm PB, Lee JY. Endoscopic approaches to brainstem cavernous malformations: Case series and review of the literature. Surg Neurol Int. 2015; 6:68.)

■ Commentary

We presented a case of a medullary cavernoma resected via an endoscopic approach. This approach allowed for resection of the lesion without brain retraction and without resection or transection of normal brain tissue. Further advantages of the endoscopic technique include the ability to limit the amount of bone removed, because the endoscope can be used with less operative exposure than the traditional operating microscope. Furthermore, the operative cavity is better illuminated by the endoscope because its light source is located closer to the operative field. Finally, a more panoramic view can be obtained with the endoscope.

We presented the endoscopic endonasal transclival resection of a medullary cavernoma to showcase an alternative approach to the treatment of a lesion similar to the lesion presented in the first section of this chapter. The far-lateral approach described in the beginning of this chapter often involves drilling the occipital condyle, which requires significant experience to obtain the perfect surgical exposure. Remove too much of the condyle, and one could destabilize the atlanto-occipital joint; remove too little, and exposure to the lesion might be insufficient and the operation compromised. Furthermore, the dissection around the occipital condyle, not to mention the drilling of it, can risk injury to the nearby vertebral artery. With the endoscopic endonasal transclival approach, these risks are eliminated.

Endoscopic techniques are not limited to the resection of medullary lesions and can be applied to lesions throughout the brainstem. These techniques are especially valuable for lesions that present to the ventral surface of the brainstem, from the pons to the cervicomedullary junction, especially since access to these areas are particularly difficult with microscopic techniques. For lesions that present to the surface more laterally, other endoscopic approaches may be employed. We have successfully resected a lateral midbrain cavernoma via an endoscopic supracerebellar infratentorial approach. For this approach, we performed a retrosigmoid craniotomy and advanced the endoscope along the undersurface of the tentorium. The operation was performed with a pneumatic arm holding the endoscope in place, as this allowed the surgeon to work with both hands. For a lateral pontine cavernoma, a standard retrosigmoid approach can be employed. Both the endoscope and traditional operating microscope may be used for these approaches, but we find that the aforementioned advantages of the endoscope argue for its utilization.

While the endoscopic technique affords many advantages, it is admittedly limited by lack of stereoscopy. Due to the lack of depth perception, and working through narrow and deep operative corridors, these cases can be quite challenging. In consideration of these limitations, endoscopic resection of brainstem lesions requires considerable experience with endoscopic techniques. While the learning curve can be steep, we believe that the advantages of the endoscopic technique outweigh the disadvantages, particularly for lesions that present ventrally.

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27 Cerebellomedullary Fissure

Daniel R. Felbaum and Walter C. Jean

Keywords: inferior cerebellar peduncle, subtonsillar approach, tela choroidea, fourth ventricle, lateral recess, tonsillobiventral fissure, supratonsillar approach

■ Clinical Presentation

A 61-year-old presented with the chief complaint of difficulty walking and frequent falls. He said that for the preceding 3 months, he felt like he was constantly being pulled toward the left whenever he walked and was unable to keep his balance. He also experienced blurry vision, but it was only because of 1 week of nausea and vomiting that he finally sought medical attention. He was a former smoker and a recent chest X-ray showed suspicious, albeit small, lung nodule. On examination, his gait was unsteady but he had no other deficits. His magnetic resonance imaging (MRI) showed a sizable cerebellar mass, which enhanced with gadolinium (**Fig. 27.1**). A body computed tomography (CT) confirmed a singular lung nodule.

Questions

1. What is the most common mass lesion in the cerebellum in an adult patient?
2. What part of the cerebellum does the tumor involve?
3. Is there any clue on the MRI image pointing to the origin of the tumor?

■ Diagnosis and Assessment

The most common cerebellar tumor in an adult is a metastasis. This is certainly at the top of the differential diagnosis for this patient, given his smoking history and the nodule found in his lung. The diagnostic considerations would have been a bit more complex had the suspicion for cancer been negligible in him. The most common *primary* cerebellar tumor in an adult is a hemangioblastoma. These tumors do not necessarily occur in patients with von Hippel–Lindau disease, and though they are most commonly cystic, they can rarely appear as a solid tumor as well. A choroid plexus papilloma must also be considered. These tumors originate in the choroid plexus of the fourth ventricle and avidly enhance with gadolinium. In **Fig. 27.1**, one can see that the inferior aspect of the tumor in this patient does in fact involve the lateral recess of the fourth ventricle, which naturally brings into consideration the involvement of the choroid plexus in this area.

Had the cancer risk in this patient been small, it would have been critical to consider both a hemangioblastoma and choroid plexus papilloma because both these tumor types are extremely vascular, and would require angiographic study and embolization of the arterial feeders prior to surgical intervention. However, given this patient's history and clinical data, the most likely diagnosis for his cerebellar tumor was a metastasis.

A biopsy of the lung lesion would yield the histopathological diagnosis of the patient's disease. Had this patient had a

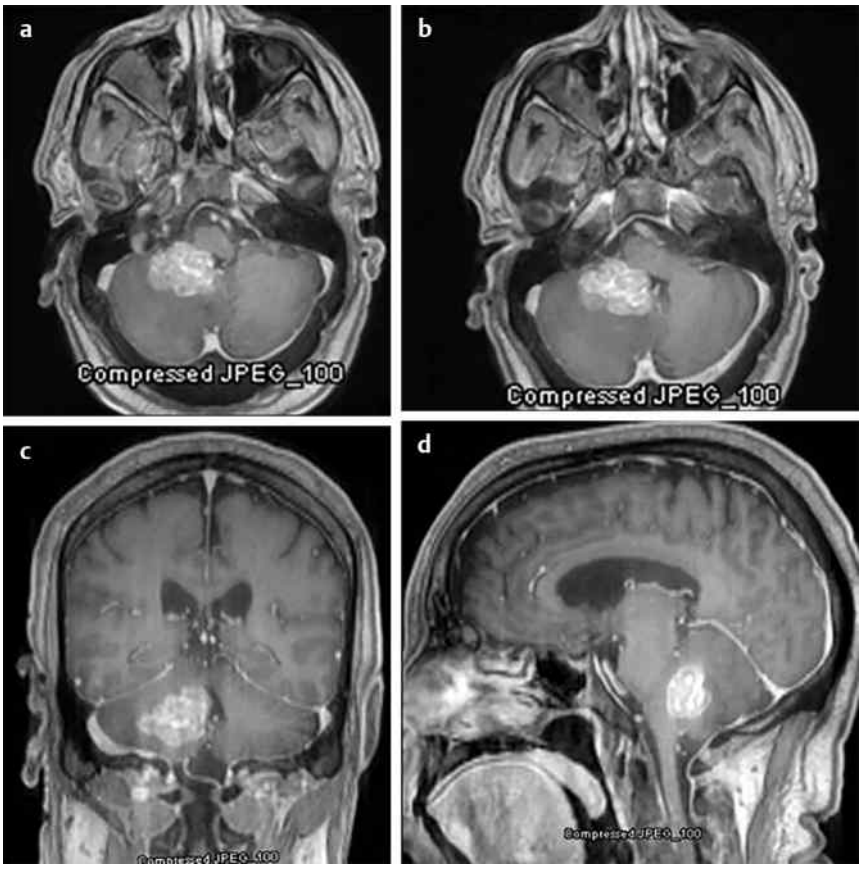


Fig. 27.1 Preoperative MRI T1-weighted images with gadolinium. (a, b) Axial, **(c)** coronal, and **(d)** sagittal images showing a tumor of the right cerebellomedullary fissure which extends superiorly into the middle cerebellar peduncle, medially into the fourth ventricle, and laterally to the foramen of Luschka.

multitude of brain lesions, too many to resect, treatable only with radiation, the biopsy of the lung lesion followed by whole brain radiation would have been the most rational strategy. As our patient only had one brain lesion, lung biopsy followed by radiosurgery is a legitimate consideration as well. However, his cerebellar tumor was on the border of being too large for radiosurgery to be effective, and given its location, there was a risk that postradiosurgery swelling would lead to obstruction of the fourth ventricle. Hydrocephalus in this setting could happen suddenly and progress rapidly. For these reasons, surgical resection of the cerebellar tumor was chosen, with the goal of complete resection, and histopathological confirmation.

■ Anatomical and Therapeutic Considerations

Anatomical examination showed that the inferior pole of the tumor filled the lateral recess of the fourth ventricle at the cerebellomedullary fissure (CMF). The tumor extended laterally to the foramen of Luschka and superiorly to just beyond the midpoint of the body of the fourth ventricle (**Fig. 27.1**). For tumors of the lower lateral cerebellum, such as the one in our patient, traditional approach options include the lateral-to-medial trajectory from a transcondylar route, or a medial-to-lateral trajectory through a transvermian or transcerebellar route (**Fig. 27.2**). The transcondylar approach would pose significant challenges since this tumor has a large midline component. The reach would be very far, and the angle suboptimal. The alternative of going through the vermis or cerebellar hemisphere would require destruction of a significant amount of normal cerebellar tissue, accompanied by inherent risks such as cerebellar mutism.

If both the lateral-to-medial and medial-to-lateral trajectories have serious weaknesses, then one must consider trajectories in the coronal and sagittal planes. In fact, in our patient, the inferior-to-superior trajectory held the key to success. The tumor originated at the lateral recess of the fourth ventricle and extended superiorly. As such, the natural cleavage plan of the CMF can be used to the patient's advantage (**Fig. 27.2**). Reaching the tumor through the subtonsillar corridor would offer excellent exposure to the tumor without sacrificing normal cerebellar tissue. While using an inferior-to-superior resection trajectory, there is always concern that a tumor extends too far superiorly to be fully removed. The tumor in our patient stopped at the level of the fastigium, where the inferior and superior medullary velum joins. In other words, the tumor stopped precisely at the superior limit for the subtonsillar approach.

The cleft between the tonsils and medulla is the CMF. Its midline limits are the fastigium superiorly and the cisterna magna inferiorly. From the midline, the fissure extends laterally to the cerebellomedullary and cerebellopontine cisterns. A vein traverses the CMF, and is an anatomical constant within the fissure. It serves as an excellent marker delineating the CMF. It is named plainly as the vein of the cerebellomedullary fissure (vCMF) and drains laterally into the superior petrosal sinus. The telovelotonsillar segment of the posterior inferior cerebellar artery (PICA) runs variably either within the fissure, or just inferior to it. This artery must be mobilized and protected as the fissure is exposed.

The floor of the fissure is equivalent to the roof of the fourth ventricle at its lateral recess (**Fig. 27.3**). This structure consists mainly of the tela choroidea which carries the vascular supply to the choroid plexus. Incision of the tela choroidea along the length of the CMF opens the lateral recess of the fourth ventricle, and allows the mobilization of the ipsilateral cerebellar tonsil dorsolaterally (**Fig. 27.3, Fig. 27.4**). This is the key maneuver to fully expose the fissure and visualize tumors in this region. At the lateral extreme of the fissure, the tela choroidea forms part of the foramen of Luschka. For surgery around this area, it is critical to know that the glossopharyngeal and vagus nerves enter the brainstem just ventral to the foramen (**Fig. 27.5**). For tumors of the CMF that extend to the foramen, such as in our patient, it is critical to appreciate that the lower cranial nerves can be disrupted by both tumor invasion and surgical manipulation. Hence, assessment of the vocal cords should be considered preoperatively. Intraoperative neuromonitoring of the lower cranial nerves with a wired endotracheal tube can aid the surgical dissection.

Questions

1. What are the limits of the CMF?
2. What artery commonly blocks the exposure of this fissure, and might be injured during its mobilization?
3. What anatomical structure must be incised to fully expose the fissure?
4. What nerves are near the lateral extreme of the fissure and what precautions should be taken if the tumor reaches this area?

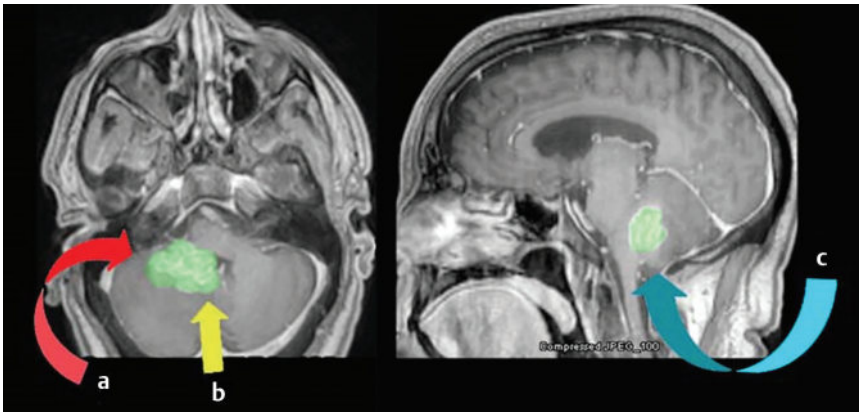


Fig. 27.2 Options for surgical approach for tumor resection. Conventional approach options include **(a)** a lateral-to-medial trajectory from a far-lateral transcondylar approach, which would pose challenges to reach the midline portion of the tumor, or **(b)** a medial-to-lateral trajectory from a transvermian/transcerebellar approach, which would entail reaching the tumor by going through significant amount of normal tissue. The inferior-to-superior approach through the subtonsillar corridor shown in **(c)** was chosen because it offers the best exposure of the tumor with the least tissue destruction.

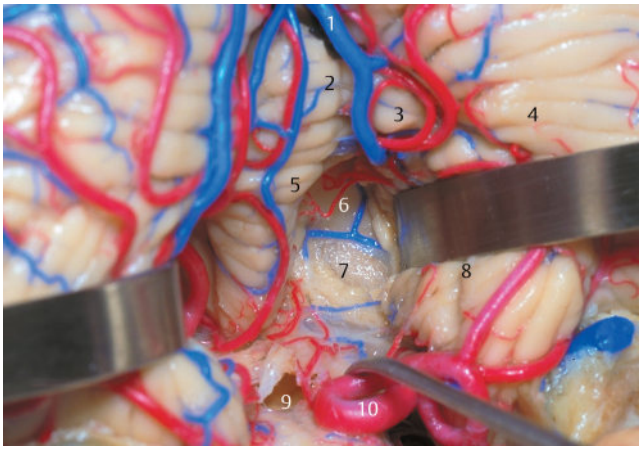


Fig. 27.3 Cadaveric dissection of the cerebellomedullary fissure. This image illustrates the subtonsillar approach to the lateral recess of the fourth ventricle. The right tonsil has been "freed" by dissecting the spaces around it (medial, anterior, and superior spaces) and retracted laterally. 1, inferior vermian vein; 2, pyramid; 3, copula pyramidis; 4, biventral lobule; 5, uvula; 6, inferior medullary velum; 7, tela choroidea; 8, tonsil; 9, foramen of Magendie; 10, PICA. Incision along the tela choroidea (7) allows the mobilization of the tonsil in a dorsal lateral direction to open the cerebellomedullary fissure.

■ Description of the Technique

The subtonsillar approach was chosen for this operation. The patient was placed in lateral decubitus position with the ipsilateral side up. The head was affixed in three-point fixation and the neck was flexed and tilted slightly toward the floor to provide maximal access to the area in the lateral suboccipital region. The mastoid prominence was kept elevated and great care was taken to protect the contralateral arm (see **The Three Approach Elements**).

A J-shaped incision was used, with the long limb in the midline and the "hook" curving toward the mastoid tip. The muscle layers were kept intact and reflected with the skin as a single mass in the inferolateral direction. A suboccipital craniotomy incorporating the foramen magnum was performed. The extent of bony removal crossed the midline on the contralateral side approximately 1 cm. Laterally on the ipsilateral side, the bone opening extended close to, but short of reaching the sigmoid sinus. The lateral lip of the foramen magnum was removed up to the occipital condyle (**Fig. 27.6**). A C1 laminectomy facilitated the dural opening.

The dura was opened in the standard Y-shaped manner (see **Operative Setup**). Under microscopic visualization, the PICA was carefully identified and mobilized away from the area of the foramen of Magendie (**Fig. 27.7**). This allowed surgical access to the CMF. The tela choroidea was identified as a visible but thin film of tissue surrounding the foramen of Magendie, and it was at the

The Three Approach Elements

Corridor: subtonsillar

Craniotomy: suboccipital

Modifiers: split CMF

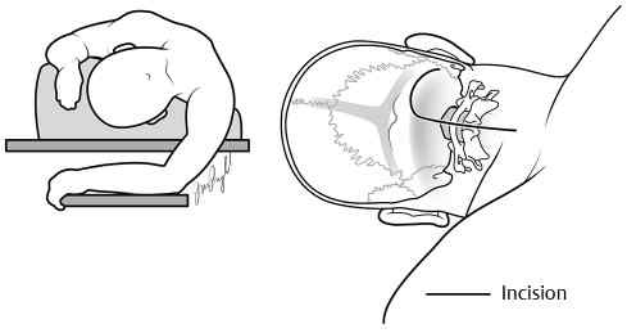
Operative Setup

Position: lateral, ipsilateral side up

Incision: J shape, hooked toward the mastoid tip

Bone Opening: lateral suboccipital, reaching occipital condyle

Durotomy: "Y"-shaped over the exposed dura



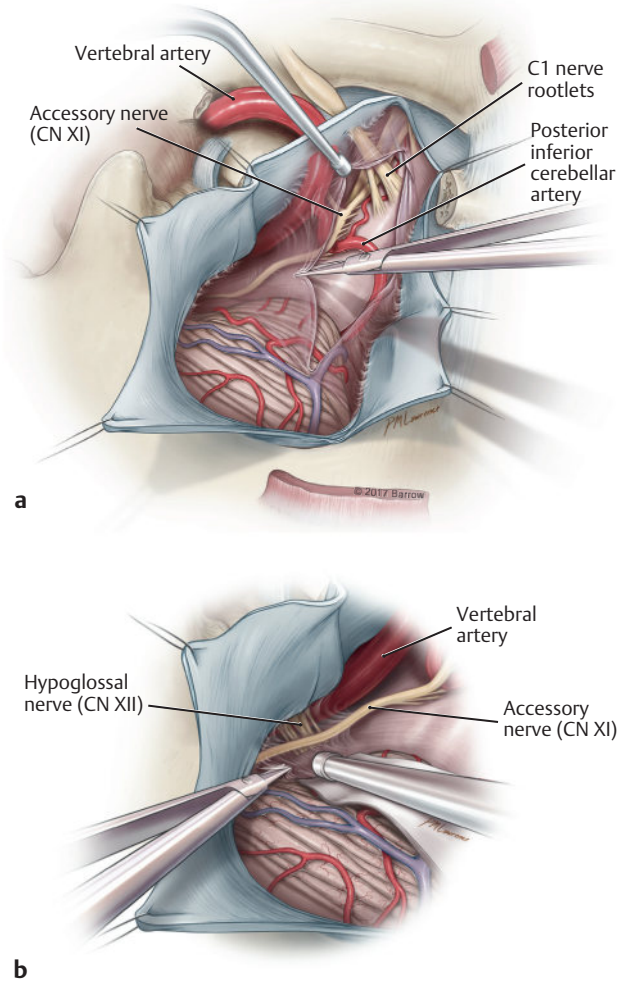


Fig. 27.4 Surgical dissection of the cerebellomedullary fissure. **(a)** The arachnoid membrane over the cisterna magna and the lateral cerebellomedullary cistern is opened. The V4 segment of the vertebral artery courses laterally to the accessory nerve (CN XI) and in front of the hypoglossal rootlets, where the posterior inferior cerebellar artery arises. **(b)** The pontine surface can be approached by dividing the arachnoid membrane covering the cerebellopontine angle cistern. Continuing the dissection ventrally leads to the premedullary cistern, providing access to the anterolateral (preolivary) sulcus and the olivary zone. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

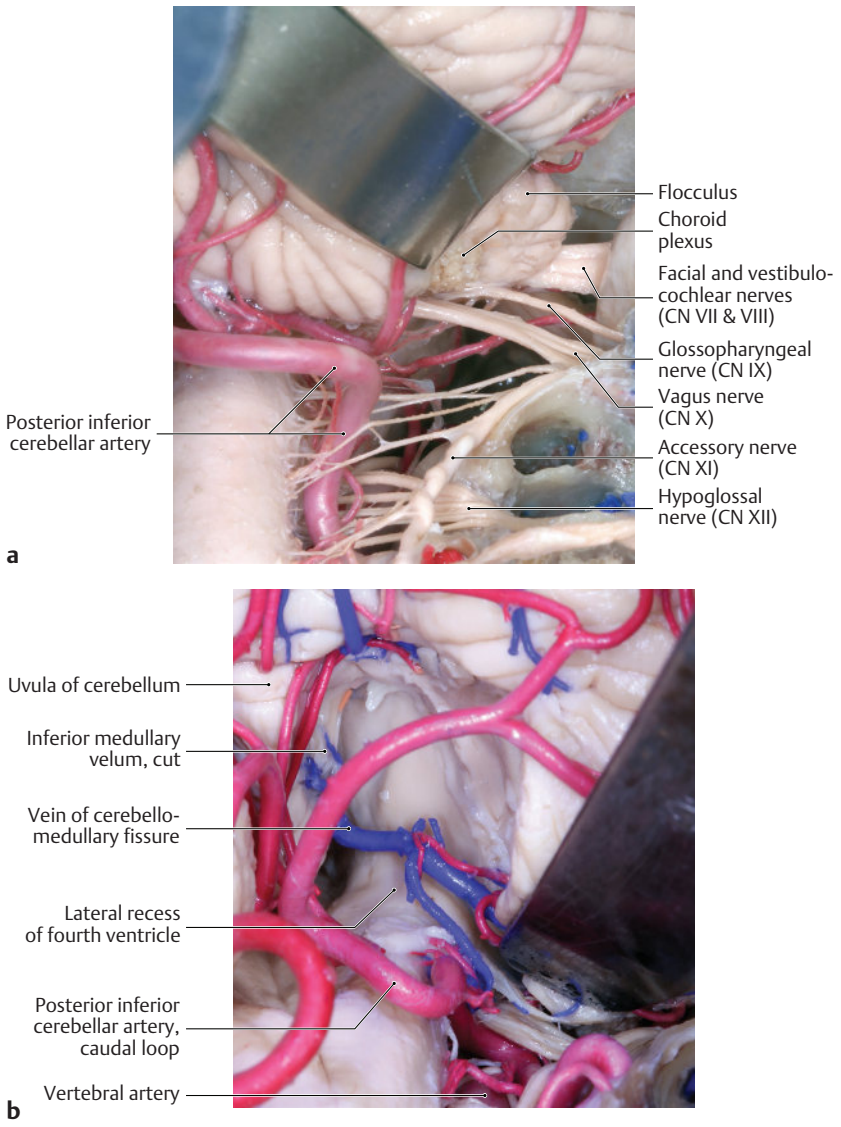


Fig. 27.5 **(a)** Dorsolateral view of the cerebellomedullary junction shows the lower cranial nerves (glossopharyngeal [CN IX], vagus [CN X], accessory [CN XI], and hypoglossal [CN XII] nerves) and their relationship to the vertebral artery and its branches. **(b)** Dorsal view of the telovelar junction and associated vessels. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

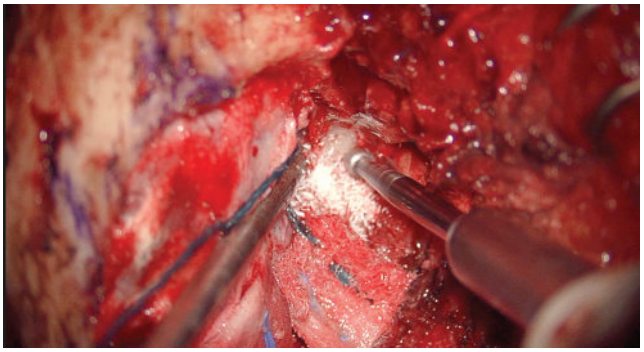


Fig. 27.6 **View at the foramen magnum**. The right lateral edge of the foramen magnum was removed to gain sufficient exposure to the subtonsillar region.

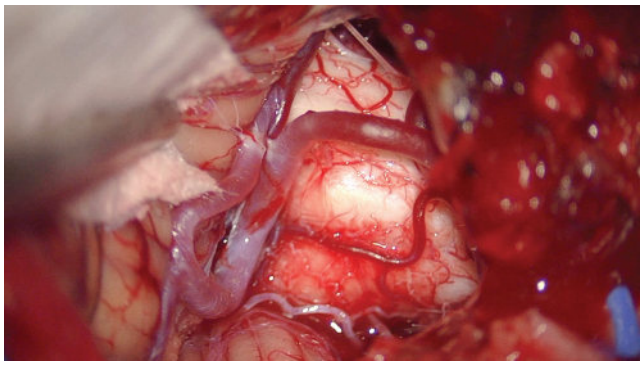


Fig. 27.7 **Posterior inferior cerebellar artery (PICA)**. With the instrument on the right cerebellar tonsil elevating it, the PICA is seen blocking access to the cerebellomedullary fissure (CMF). The artery must be carefully mobilized away to gain exposure to the CMF.

foramen that it was first incised (**Fig. 27.8**). The exposure of the fissure then followed the vCMF from a medial to lateral direction. As dissection continues, the tela choroidea was stepwise exposed and incised (**Fig. 27.9**). As a result, the lateral recess of the fourth ventricle was fully exposed, and the tumor therein was visualized (**Fig. 27.10**).

The tumor was debulked at this point, but it proved to be extremely vascular. As such, arterial feeders had to be found

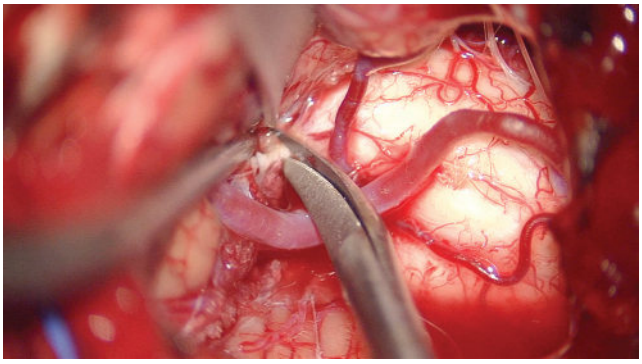


Fig. 27.8 Incising the tela choroidea. Micro-scissors were used to cut the tela choroidea covering the right dorsal aspect of the foramen of Magendie. This was the initial step for accessing the lateral recess of the fourth ventricle in the subtonsillar region.

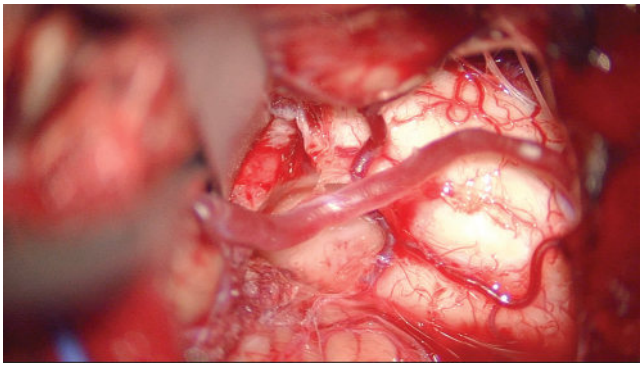


Fig. 27.9 Tumor exposure in the cerebellomedullary fissure (CMF). After the incision in the tela choroidea over the foramen of Magendie, the most medial portion of the CMF has been opened, revealing the inferomedial edge of the tumor under the retractor.

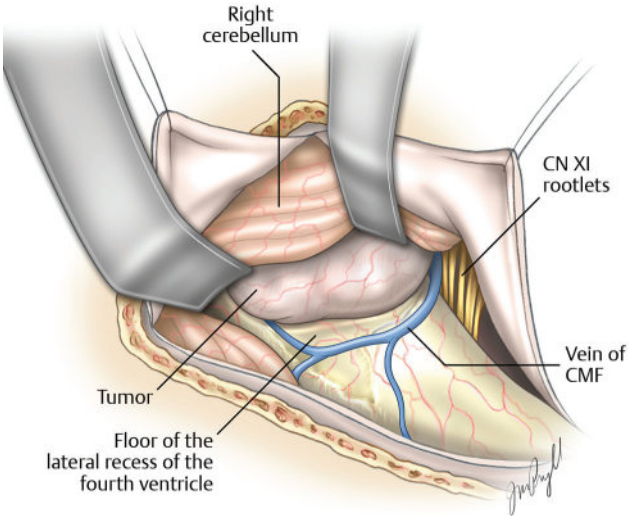


Fig. 27.10 Tumor exposure. Illustration of the tumor fully exposed in the cerebellomedullary fissure, after the tela choroidea was opened laterally toward the foramen of Luschka.

and eliminated early. Some piecemeal resection allowed the surgeon to reach the arterial supply safely. Dissection of the rest of the tumor started at the floor of the fourth ventricle, moved laterally toward the foramen of Luschka, with great care to preserve the lower cranial nerves, then proceeded back medially along the top surface of the tumor. Where appropriate, tumor aspirators were used to facilitate tumor removal until it was complete (**Video 27.1**).

For closure, the dura was reapproximated with sutures and this closure was augmented with a dural substitute in an onlay manner. The craniotomy bone flap was replaced and the rest of the soft tissue closed. The skin was closed with a running nylon stitch to obtain watertight closure. The postoperative MRI showed a complete resection (**Fig. 27.11**).

Surgical Pearls

1. For surgeons that are unfamiliar with the cerebellomedullary fissure dissection, it is easy to get "lost" and neuronavigation in the lateral position might compound the problem. The vCMF is an excellent "marker" for the fissure as it is usually large and invariable in its location. Using it as a guide for the dissection can be helpful for orientation.
2. The telovelotonsillar segment of the PICA almost uniformly blocks the cerebellomedullary fissure at its inferior edge. This artery can be mobilized away by carefully cutting the arachnoid attachments to the brainstem and cerebellar surfaces. Once mobilized, it is critical to ensure that the artery is well-padded, without kinks while it is held away from its natural position at the CMF.
3. If the tumor has a significantly large intraparenchymal component within the cerebellar hemisphere, the natural advantage of using the CMF would be lost, and a transcerebellar route may be more practical.

■ Aftercare

After the operation, the patient had new deficits of right-sided dysmetria and dysfunction of the right vocal cord. He also experienced rotatory nystagmus which fortunately improved rapidly. Histopathological examination revealed a small cell carcinoma entangled with choroid plexus and cerebellar tissue, and given the patient's smoking history and lung nodule, all the findings were consistent with a metastatic lung cancer.

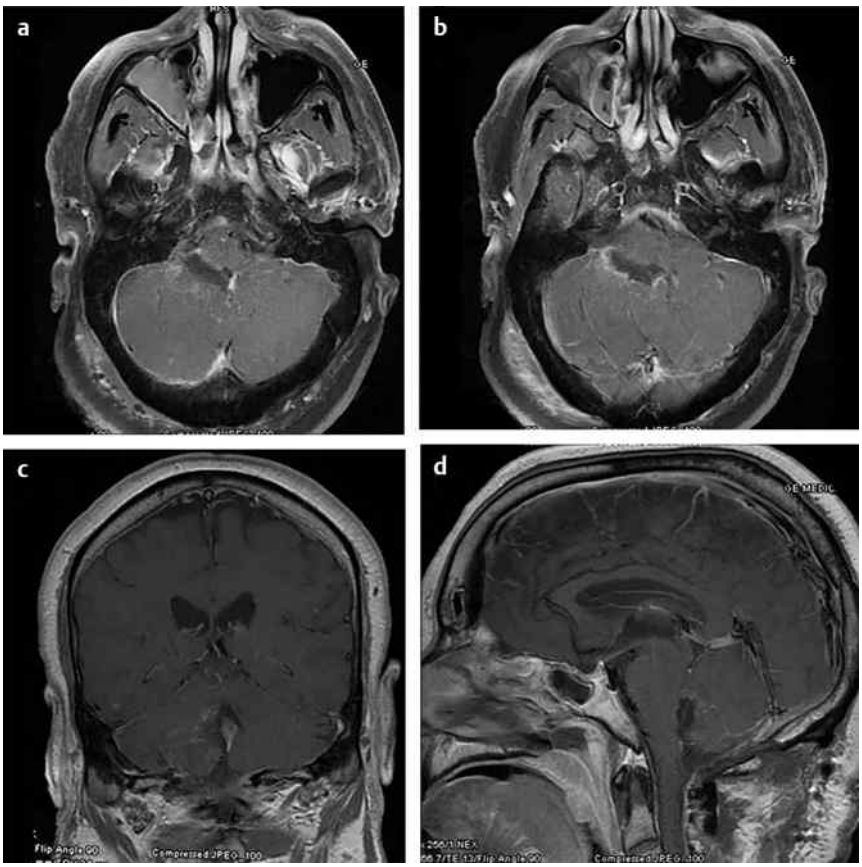


Fig. 27.11 Postoperative MRI T1-weighted images with gadolinium. (a, b) Axial, **(c)** coronal, and **(d)** sagittal images showing a total resection of the tumor. Histopathological identification was consistent with a small cell carcinoma. Given the patient's lung nodule, this was most likely from a lung primary.

■ Possible Complications and Associated Management

Cerebellar injury, hydrocephalus, and cranial nerve dysfunction are the main problems that can complicate the subtonsillar approach to tumor resection in the CMF. In order to minimize excessive cerebellar retraction, the bony opening must be sufficient to allow for mobilization of the tonsil. To optimize the trajectory, the dorsomedial portion of the occipital condyle may need to be thinned, making the operation a far lateral partial transcondylar approach.

The operation necessitates the lateral recess of the fourth ventricle be wide open for surgical exposure. Debris from the dissection and tumor removal can travel through the cerebrospinal fluid (CSF) and lead to postoperative hydrocephalus. To minimize this risk, cottonoids can be used to block the cerebral aqueduct at the superior pole of the surgical field. Although debris can still travel through the CSF, its backflow into the third and lateral ventricle will at least be kept to a minimum with this precaution.

Dissection around tumors in this region is fraught with peril, as the tumor is surrounded by delicate structures of the medulla ventromedially, and by the lower cranial nerves laterally. For tumors large enough to extend laterally to the foramen of Luschka, the glossopharyngeal and vagus nerves will not function properly after surgery. These nerves exit the brainstem directly ventral to the foramen of Luschka, and whether by surgical manipulation or by tumor entanglement, they will almost certainly be compromised at least temporarily. Intraoperative monitoring of these nerves will not prevent their injury, but may help guide postoperative management. But even if monitoring data indicate that the lower cranial nerves should be intact, postoperative extubation must be done with extreme care. The patient must not be given an oral diet until swallowing function can be studied in detail after the surgery. If the study is equivocal, a liberal consideration of percutaneous feeding tubes placement can minimize the incidence of aspiration pneumonitis.

Perspective

Kyle Mueller and Walter C. Jean

■ Introduction

Although the superior limit of the subtonsillar approach is the inferior cerebellar peduncle (ICP), it is tempting to pursue every lesion that involves the cerebellar peduncle with this technique because the separation of the tissue planes in the CMF is so "natural," and can be accomplished with so little damage to the cerebellum. However, for lesions in the ICP that do not descend into the CMF, but extend further superiorly into the *middle* cerebellar peduncle (MCP), it may be wise to admit that the subtonsillar route simply does not provide adequate reach and resort to alternative approaches. Fortunately, for lesions that are centered more superiorly, there is a variation of the approach, above the tonsil, that can provide satisfactory exposure.

■ Case Presentation

A 68-year-old man of African descent was in his usual state of health until one day when he suddenly felt ill and nauseous, and returned home from work early. The next day, his family brought him to the emergency department for right facial numbness and difficulty walking. He fell to the right when he tried to walk. His medical history was significant for previous strokes. His physical examination showed right-sided dysmetria, numbness on the right face (V_{1-3}), and right facial palsy (House–Brackmann 2). MRI of the brain was performed and it showed a lobulated mass in the right MCP with peripheral enhancement (**Fig. 27.12a**). The lesion also showed restricted diffusion on diffusion weighted images (DWI). The rest of the study was only significant for evidence of the patient's previous ischemic strokes, one of which involved the contralateral cerebellum.

■ Anatomical and Therapeutic Considerations

In an elderly man, with a ring-enhancing cerebellar lesion, the first thought was naturally that the lesion represented a metastasis from systemic cancer. For this reason, CT of the chest, abdomen, and pelvis was performed soon after his admission, but it did not reveal any evidence of systemic cancer. With this patient's history and the restriction on DWI, one concern was that he suffered yet another stroke, but the enhancement pattern made this somewhat unlikely. An infectious process such as an abscess would also restrict on DWI, and as such, given that there was no mass effect, a lumbar puncture was performed. The results showed that the CSF glucose was 10 mg/dL, adding evidence that the lesion was most likely an abscess. An intracranial abscess can be a life-threatening condition, especially if it ruptured into the ventricular system. By its MRI appearance, this abscess had reached late capsular phase, and because of this, we decided to surgically remove the abscess.

The lesion in this patient was located mainly in the MCP, and it had no exposed surface in the CMF or the lateral recess of the fourth ventricle. Therefore, neither the subtonsillar or transvermian approach would adequately access this lesion. In order to avoid traversing large volumes of healthy cerebellum, the supratonsillar approach can be utilized by approaching the lesion through the tonsillobiventral fissure (**Fig. 27.13**). This fissure is a secondary fissure that separates the cerebellar tonsil from the biventral lobule (**Fig. 27.14**). Even though it is somewhat obscure because of its narrow width, identification of the fissure is a key step in the supratonsillar approach. The orientation of the folia on the tonsil is orthogonal to that on the biventral lobule, and this unique geometrical feature can help locate the fissure. In addition, terminal branches of the PICA dive into the fissure, and as long as one does not injure these branches, separation of the fissure can start by following them. At the base of the tonsillobiventral fissure is the tonsillar peduncle, a white matter tract that connects the tonsil with the cerebellum. This tract will merge with the ICP, and this allows the surgeon access to lesions here as well as in the MCP.

■ Description of the Technique

The patient was induced under general anesthesia and positioned to allow us to look down the axis of the tonsillar peduncle.

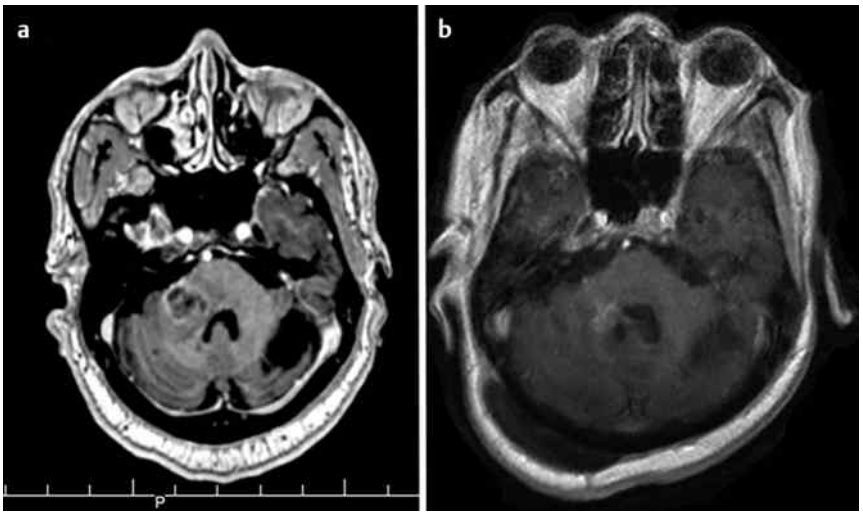


Fig. 27.12 Pre- and postoperative MRI. **(a)** MRI with gadolinium showing an old infarct in the left cerebellar hemisphere and a rim-enhancing lesion in the right middle cerebellar peduncle. **(b)** MRI 2 months after surgery showing near-resolution of the lesion.

This involved a three-quarter prone position, with head rotated about 30 degrees away from the side of the lesion, plus laterally flexed 30 degrees toward the floor. The head position was fixed and neuronavigation was registered. An "L-shaped" incision was made with the short limb just inferior but parallel to the transverse sinus, and the long limb down to the C2. A craniotomy was performed that extended from the right transverse sinus down to the foramen magnum. A right-sided hemilaminectomy of (C1) was also performed.

The dura was then opened in a Y-shaped manner and CSF was released from the cisterna magna. The tonsillobiventral fissure was identified using the landmarks described above and careful sharp arachnoid dissection was performed, taking care to preserve PICA branches (**Fig. 27.15**). The fissure split guided the direction of dissection with additional assistance from neuronavigation.

In contrast to the subtonsillar approach in which the surgeon works on the inferior medial side of the tonsils, the supratonsillar approach focuses on the superior lateral aspect of the tonsil with the retractor holding the biventral lobule superolaterally. Alternatively, a retractor can be used to move the tonsil medially and inferiorly. After dissection of the tonsillobiventral fissure, the navigation was utilized to optimize the shortest trajectory to the abscess. A small incision was made in the white matter tract and the abscess was visualized. Cultures were

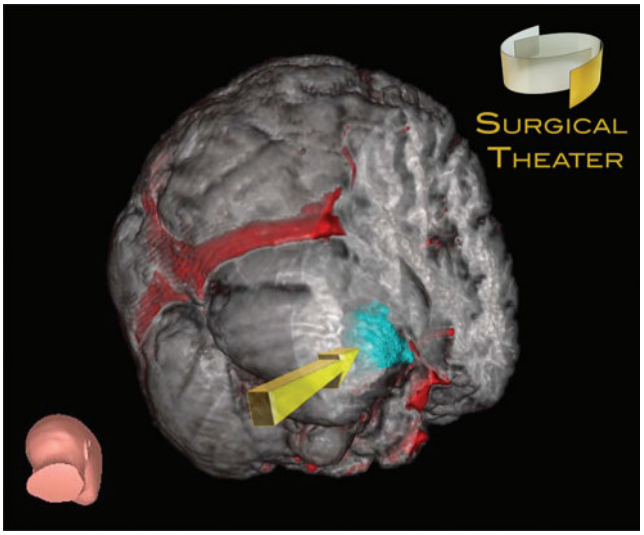


Fig. 27.13 Three-dimensional rendering in three-quarter prone position. Surgical Theater rendering of the patient's brain in three dimensions with yellow arrow showing the supratonsillar trajectory to the target lesion in the right middle cerebellar peduncle. Inset: orientation of the head.

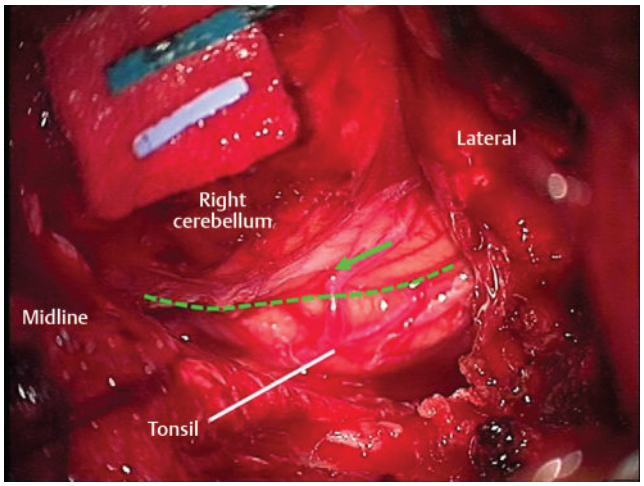


Fig. 27.15 Intraoperative view. View under the microscope of the right cerebellar hemisphere. The green line indicates the tonsillobiventral fissure just prior to surgical dissection. The green arrow indicates a posterior inferior cerebellar artery branch diving into the fissure.

Fig. 27.14 The tonsillobiventral fissure. This is a dorsal view of the suboccipital surface of the cerebellum. The yellow line denotes the tonsillobiventral fissure superior to the cerebellar tonsil. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

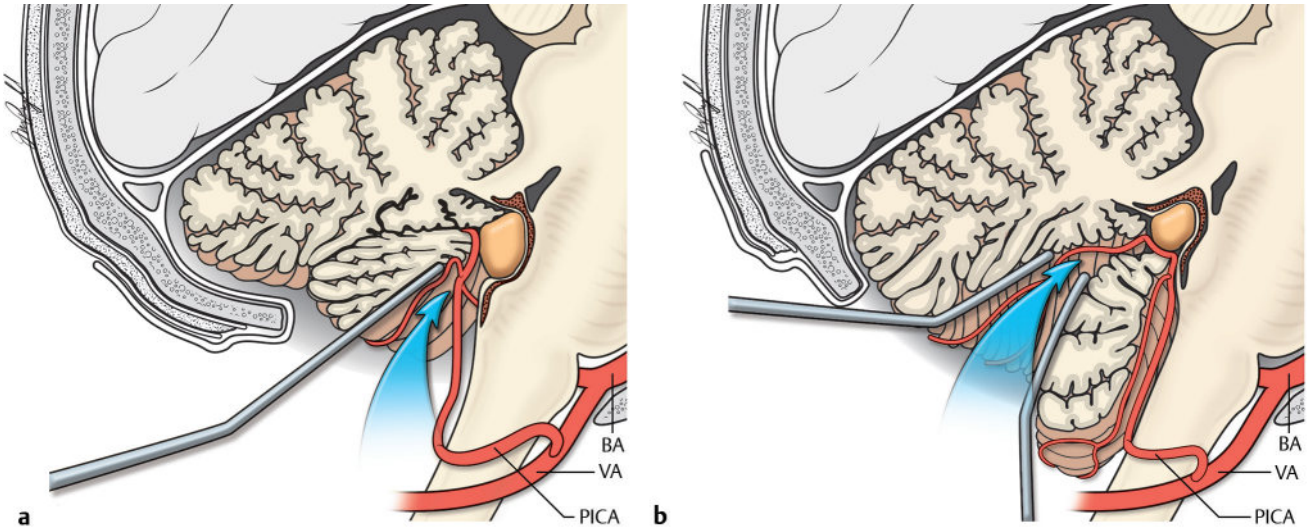


Fig. 27.16 Comparison of the subtonsillar and supratonsillar approaches. (a) The subtonsillar approach opens the cerebellomedullary fissure and targets lesions in the lateral recess of the fourth ventricle. **(b)** The supratonsillar approach opens the tonsillobiventral fissure and targets lesions more superiorly, including the inferior cerebellar peduncle. BA, basilar artery; PICA, posterior inferior cerebellar artery; VA, vertebral artery.

sent of the purulent material, and the rest of the abscess was removed with microsurgical technique. Thereafter, hemostasis and dural closure were performed and the rest of the closure was done in standard fashion.

■ Aftercare

The patient was extubated and transferred to the neurointensive care unit for close monitoring. Routine postoperative imaging showed complete evacuation of the abscess (**Fig. 27.12b**). Cultures indicated that the offending microorganism was Eikenella corrodens. Without additional neurological complications, the patient was started on intravenous antibiotics and transferred to rehabilitation unit. He developed hydrocephalus two months after surgery and required CSF diversion. He has since made steady recovery towards his neurologic baseline.

■ Commentary

Taking advantage of the CMF to access the fourth ventricle, the telovelar approach, if the lesion is bilateral, or the subtonsillar approach if unilateral, offers the surgeon major advantages as these approaches use the natural planes to avoid damaging healthy cerebellum. But these approaches can only reach as far as the inferior aspect of the ICP, and using them to go further superiorly is unrealistic. For the rare lesions situated in the MCP, the supratonsillar approach through the tonsillobiventral fissure is a better alternative (**Fig. 27.16**).

In addition to a more superior reach, the supratonsillar approach differs from its subtonsillar “cousin” in important ways. If one considers the brainstem as a reference axis, then the subtonsillar approach is directed horizontally, but the supratonsillar approach is vertical, as the tonsillobiventral fissure is nearly parallel to the brainstem. This fissure also naturally directs the operative trajectory more laterally than the subtonsillar approach, and this allows the surgeon to avoid the dentate nucleus and limit mutism complications, which we did not see in our patient.

Identification of the anatomical fissure is one of the most challenging aspects to dissecting the tonsillobiventral fissure. The orientation of the folia and radiation of distal PICA branches are key landmarks. Retraction on the biventral lobule is almost unavoidable during the fissure split, as pial adhesion within this fissure is tighter than the CMF. At the end of the fissure, it is often necessary to dissect through small parts of the tonsillar peduncle to access the lesion. In patients that have undergone this procedure, this does not appear to be associated with major morbidity.

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28 Torcula

Jacob Ruzevick and Manuel Ferreira Jr.

Keywords: meningioma, sinus reconstruction, superior sagittal sinus, transverse sinus

■ Case Presentation

A 72-year-old, right-handed woman presented with headache, subtle cognitive decline, and ataxia of 6 months' duration. Her examination was notable for ataxia, but her cranial nerves were normal. Specifically, her visual fields seemed largely intact, though no formal ophthalmologic exam was obtained. An initial cranial computed tomography (CT) study revealed a 7-cm, partially calcified, heterogeneous, falcotentorial-based mass extending into both the supra- and infratentorial spaces. There was significant mass effect causing displacement of the right occipital lobe and cerebellar hemisphere, with compression of the fourth ventricle causing mild ventriculomegaly (**Fig. 28.1**). Because of her headaches, increasing difficulty with walking, and the dramatic findings of her CT, she was referred to our unit for intervention.

Questions

1. What additional assessments are needed before you can plan any intervention on this patient?
2. How would this patient's age affect your decision-making on what to do for her?

■ Diagnosis and Assessment

A subsequent magnetic resonance imaging (MRI) confirmed a large, heterogeneously enhancing, dural-based mass extending both into the supra- and infratentorial spaces. It invaded the right transverse sinus, and involved the vein of Galen, the straight sinus, and the torcula Herophili. In addition, there was an acquired Chiari malformation with approximately 1 cm of tonsillar descent through the foramen magnum due to mass effect. There were peritumoral FLAIR signal changes within the brain parenchyma and intratumoral flow voids. There was minimal diffusion restriction within the tumor (**Fig. 28.2**).

The imaging characteristics of this tumor were suggestive of a meningioma, though other tumors in the differential diagnosis included metastasis, or hemangiopericytoma. A body CT scan was performed to identify any extracranial sites of disease that might suggest a dural-based metastasis, since common metastases, including breast, lung, and prostate cancers can show similar imaging characteristics such as avid enhancement and a dural tail. No systemic cancers were found.

Tumors in this location can displace or occlude normal vascular anatomy, and therefore an understanding the vascular anatomy, especially related to the dural venous sinuses, is critical to successful and safe tumor resection as in **Fig. 28.3**. While the technology for noninvasive imaging is improving, digital subtraction angiography (DSA) remains the gold standard for identifying the vascular anatomy surrounding the mass, as well as determining any feeding vessels to the tumor, which could be eliminated via preoperative embolization. For a tumor like this, the role of embolization is as dependent on the operative surgeon's preference, as the role of embolic material is on the endovascular one. Infiltrating small particles, such as polyvinyl alcohol particles, provide better tumor penetration but are associated with a higher risk of posttreatment complications, such as cranial neuropathies and hemorrhage due to occlusion of venous outflow. Current literature suggests that the overall complication rate from tumor embolization is approximately 6%. Timing of surgery following embolization also remains surgeon specific, with multiple groups suggesting 1 to 10-plus days gap between the two procedures.

In our case, the DSA showed near-complete occlusion of the right transverse sinus with normal filling of the left transverse sinus, inferior sagittal sinus, and superior sagittal sinus (SSS). The primary arterial feeder was through a transosseous branch of the occipital artery, which was successfully embolized (**Fig. 28.4**).

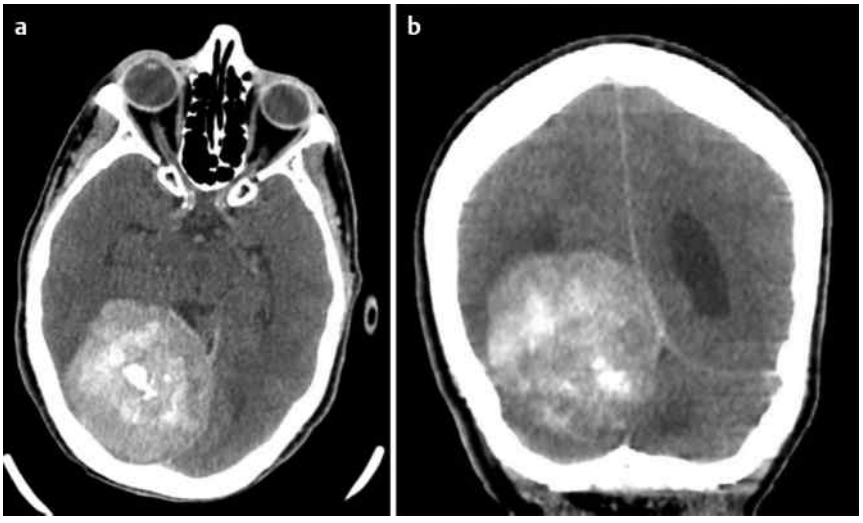


Fig. 28.1 **(a)** Axial and **(b)** coronal CT showed a 7-cm, partially calcified, heterogeneous, falcotentorial-based mass extending into both the supra- and infratentorial spaces.

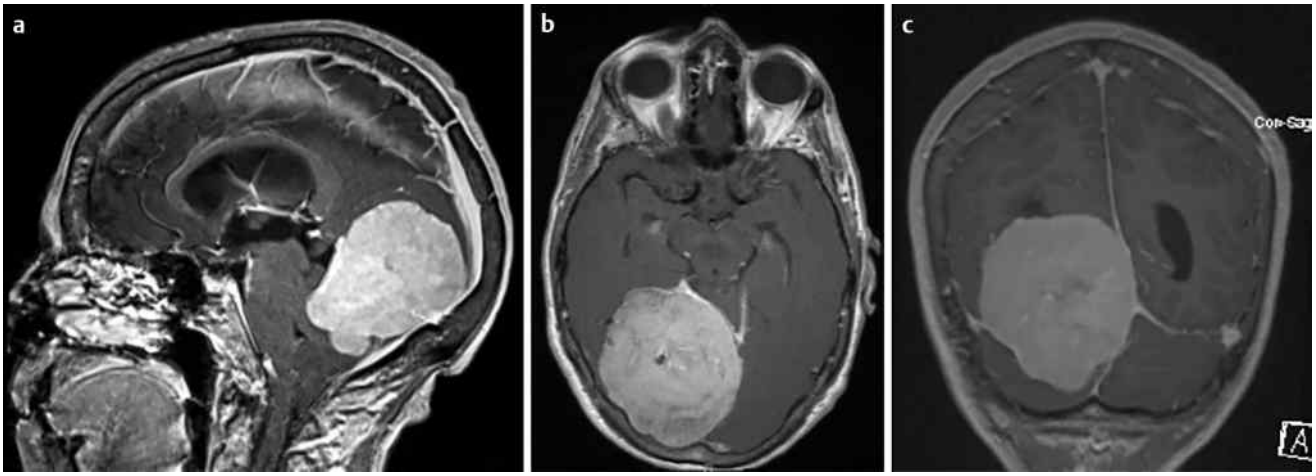


Fig. 28.2 **(a)** Sagittal, **(b)** axial, and **(c)** coronal MRI showed a heterogeneously enhancing, dural-based mass involving the tentorium and falx. The mass extended both into the supratentorial space, causing significant mass effect on the right greater than left occipital lobe, and into the posterior fossa. The posterior fossa aspect of the tumor caused mild displacement of the vein of Galen and the straight sinus.

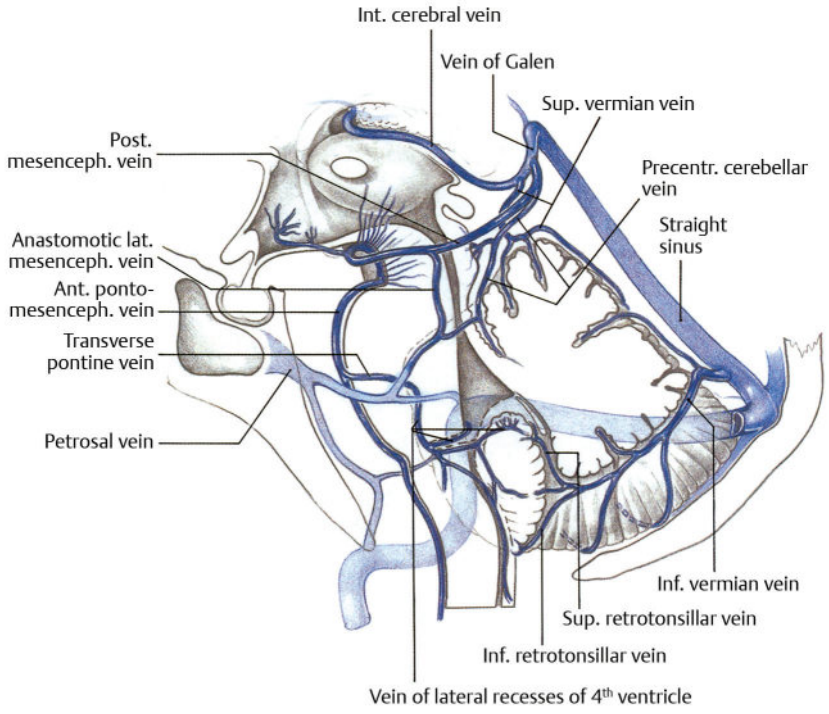


Fig. 28.3 Anatomy of the dural sinus and posterior fossa venous system. (Reproduced from Yaşargil M, Teddy P, Valavanis A, et al, eds. Microneurosurgery, Volume III A. AVM of the Brain, History, Embryology, Pathological Considerations, Hemodynamics, Diagnostic Studies, Microsurgical Anatomy. 1st ed. Thieme; 1987.)

■ Anatomical and Therapeutic Considerations

Operative intervention should be made after a thorough discussion of all risks and benefits. The patient presented here was symptomatic from this lesion, and thus operative resection was recommended to immediately remove the mass effect. In this setting, radiation can be used as an adjuvant therapy if residual tumor is present after surgery, but cannot be used as primary "stand-alone" therapy because of the tumor size. The addition of adjuvant radiation minimizes the risk of venous sinus thrombosis from removing tumor *within* the sinuses. However, of those patients treated with adjuvant radiation after subtotal resection, up to 75% experience recurrence of disease and approximately half suffer a neurological complication. This indicates to us that total resection, when safe, is the optimal management.

Options for Approach

A wide exposure is required for surgery on torcular meningiomas, mainly to have control of surrounding venous structures. For tumors that invade the dural sinus, the surgeon must decide whether to perform a *radical resection*, including removing the tumor inside the sinus followed by repair of the sinus wall, or *partial resection*, removing the tumor up to the sinus wall

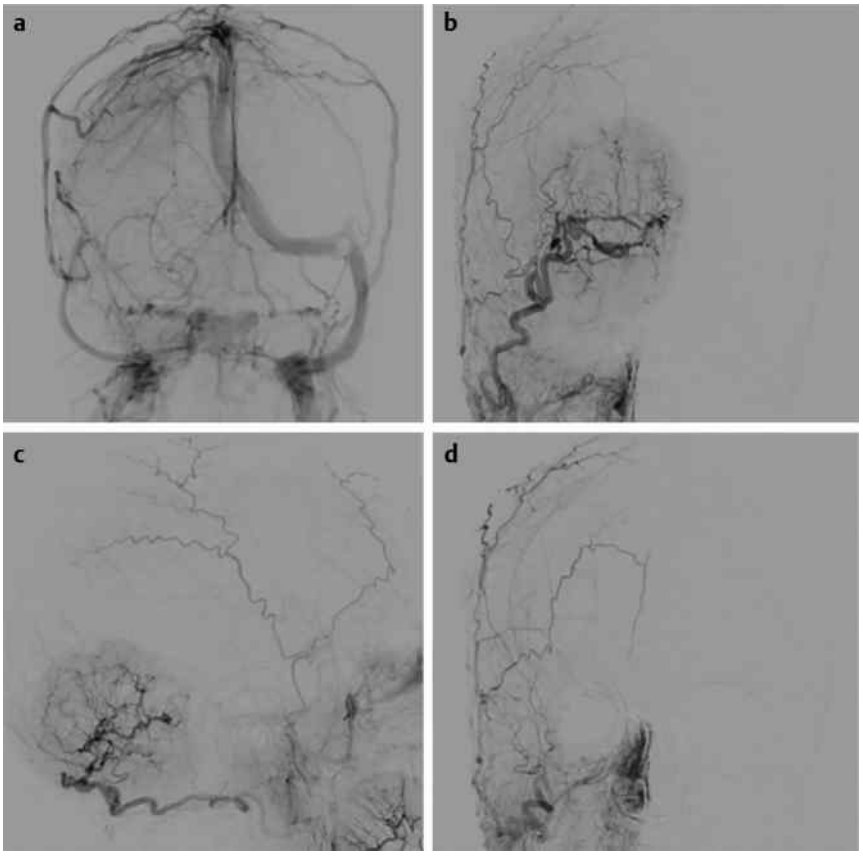


Fig. 28.4 Preoperative angiogram. **(a)** Digital subtraction angiography showed minimal filling of the right transverse sinus suggesting near complete occlusion from tumor invasion. The left transverse sinus, inferior sagittal sinus, and superior sagittal sinuses filled normally. **(b)** Anteroposterior and **(c)** lateral projections showed the primary arterial supply was through a transosseous branch of the occipital artery and imaging of the tumor showed a tumor blush consistent with meningioma. Multiple cortical vessels were displaced over the superior aspect of the tumor. **(d)** Postembolization imaging showed complete occlusion of the feeding transosseous branch of the occipital artery.

with plans for adjuvant therapy. While removal of a sinus *with the tumor* is typically safe if the sinus is completely occluded, this must be confirmed with both preoperative angiography and intraoperative Doppler ultrasound or indocyanine green angiography. When sinus resection or temporary ligation is considered, anatomical nuances must be carefully considered, including identification and preservation of venous collaterals, patency of the contralateral transverse sinus, and identification of the vein of Labbé.

Operative management of a completely occluded sinus can include either reconstruction of the invaded sinus or ligation and removal of the occluded segment (**Fig. 28.5**). Ligation should only be done if there is adequate collateral venous drainage and capacity of the contralateral transverse sinus to handle increased venous outflow. Delayed, venous-phase images from the DSA are useful to make this determination. In the torcular region, it is critical to note the insertion of the vein of Labbé as the transverse sinus can only be ligated medial to this insertion. Prior to permanent ligation, temporary clips can be placed proximal and distal to the area of luminal obstruction, while the brain is assessed for potential swelling. Testing for changes in pre- and post-clip distal venous pressure is another way to assess whether permanent ligation is safe (**Fig. 28.6**).

Operative management of a partially occluded sinus should focus on total tumor resection and restoring patency of the dural sinus. Tumor resection from a sinus with at least one wall affected is made more challenging due to the need for potential shunting during tumor resection. Our practice, however, is to allow for controlled sinus bleeding in lieu of sinus shunting. Following tumor resection from the sinus, it can be repaired with

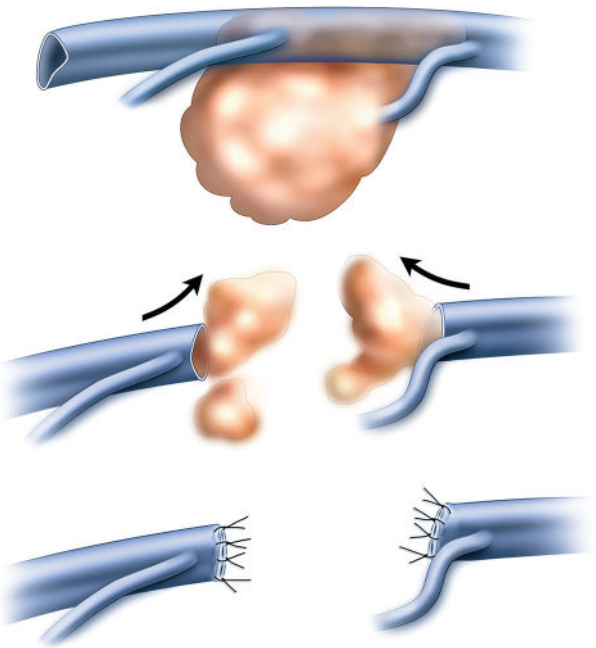


Fig. 28.5 Removal of an occluded sinus with the tumor. When an involved sinus is completely occluded by tumor, consideration can be made for removal of the sinus with the tumor. The proximal and distal ends of the sinus are then ligated, provided that collateral venous drainage is sufficient. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

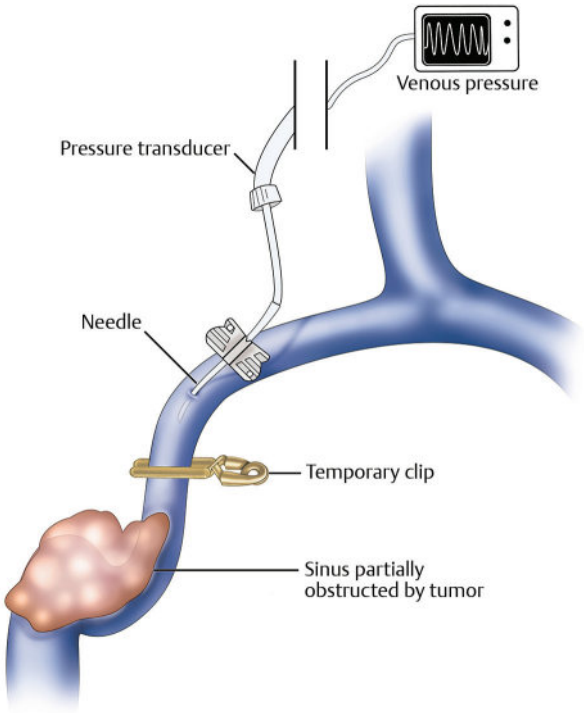


Fig. 28.6 Testing for change in pre- and post-clip venous pressure. A 20-gauge needle inserted into the sinus is connected to a pressure monitoring line to measure the intraluminal pressure. If there is no significant rise of the pressure after temporary occlusion of the sinus for 1 minute, then the sinus can be transected/ligated. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

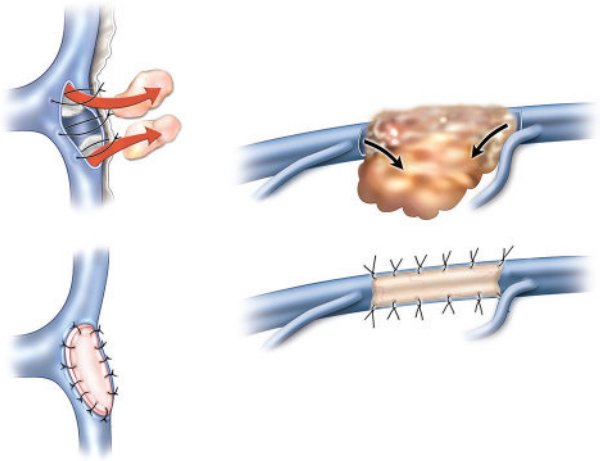


Fig. 28.7 Wall reconstruction of an involved sinus. An autologous dural patch is sutured into place after removal of tumor from a partially occluded sinus. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

primary suture, with patching, or with an autologous bypass. Patching can include either autologous material, such as dura, pericranium, fascia, or synthetic material (**Fig. 28.7**). However, a significant risk of patching is the prolapse of the patch into the lumen of the vessel with subsequent obstruction of venous flow. Synthetic bypass grafts have a high rate of thrombosis and are generally not recommended. As with the patient presented here, our preference is to primarily repair the sinus. The sinus is opened along the path of the tumor, and once the intraluminal tumor is removed, the venous flow is partially slowed with external compression of the sinus. Instead of using temporary clips or intraluminal hemostatic agents, both of which can damage the endothelium, we find that gentle compression by the surgeon's assistant is the best way to tamponade the venous bleeding during wall repair. And as the tumor is delivered out of the sinus, the sinus wall is continuously closed with suture (**Fig. 28.8**).

Approach of Choice

In our 72-year-old patient, with a nearly occluded right transverse sinus, one can make a case for partial resection followed by radiation. However, given the aforementioned high

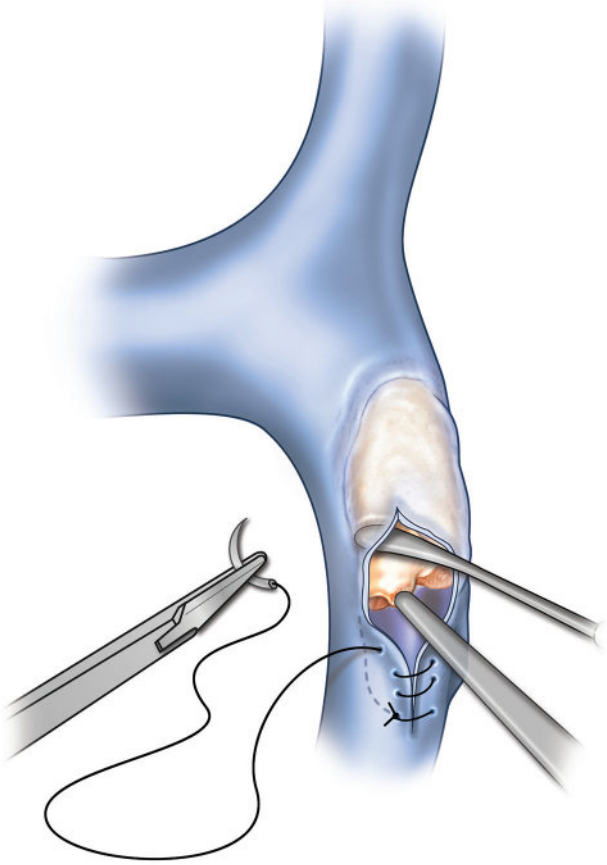


Fig. 28.8 Bleeding from the sinus after restoration of blood flow is controlled by gentle tamponade by the surgeon's assistant (not shown).

recurrence rate in patients treated this way, the unknowable possibility that this meningioma could be above WHO I with an even higher risk of recurrence, and the projected longevity of this patient without comorbidities, our plan was to open the involved sinus, and resect the tumor therein. We did not consider ligation of her transverse sinus, as in our practice, if a tumor-involved sinus is patent, even if only partially, we will attempt tumor resection from the invaded sinus. This step is always planned as the last maneuver of the operation, and all patients considered for this are placed on aspirin preoperatively.

Questions

1. How would you handle this patient's hydrocephalus before or during her operation?
2. What position would you use for her operation?
3. What precautionary steps would you take before you start her operation?

Description of Technique

After establishment of general anesthesia, a precordial Doppler was used to monitor for intraoperative air embolism. To begin, an external ventricular drain was placed at Kocher's point. The patient was then repositioned prone and the head was fixed in a Mayfield head clamp. The head was placed in a military tuck position (see **The Three Approach Elements**). Prior to incision, a 1 g/kg bolus of mannitol was infused to assist in brain relaxation. A midline vertical incision was performed and carried to the bone using monopolar cautery. Burr holes were placed straddling the SSS and the transverse sinuses on either side. The dura was then stripped to avoid laceration from the craniotome. A single bone flap incorporating the occipital and suboccipital regions was then elevated with care not to injure the sinuses. For this case, dura was opened in a stellate fashion over the occipital aspect of the tumor. An additional durotomy was made to expose the infratentorial aspect of the tumor (see **Operative Setup**).

The Three Approach Elements

Corridor: posterior

Craniotomy: occipital and suboccipital

Modifiers: open an.d repair of transverse sinus

Tumor resection was performed in stages with the first goal being to safely resect the supratentorial aspect of the tumor (**Fig. 28.9**). Veins en passage were identified and spared. In addition, tumor adherent to critical brain or vascular structures can be purposefully spared with goals of treating with adjuvant therapy. Tumor was then resected from the infratentorial space (**Fig. 28.10**). Following safe resection of all identifiable tumor, the ipsilateral transverse sinus as carefully inspected. The point of invasion by the tumor was clearly identifiable and most of the wall was normal. The sinus was then opened to resect infiltrative tumor, starting just lateral to the distal point of tumor invasion into the transverse sinus (**Fig. 28.11**). Following resection, and with the surgeon's assistant compressing the proximal sinus to control the restored venous flow, the wall was primarily closed with 5–0 Prolene suture from a distal to proximal direction (**Fig. 28.12**). Doppler

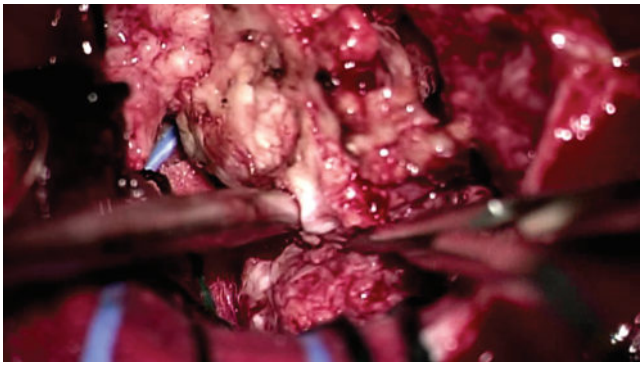


Fig. 28.9 Intraoperative view of initial resection of the occipital extension of the tumor.

Fig. 28.10 Intraoperative view of the dural entry sites for the supra- (*left*) and infratentorial (*right*) areas of tumor resection.

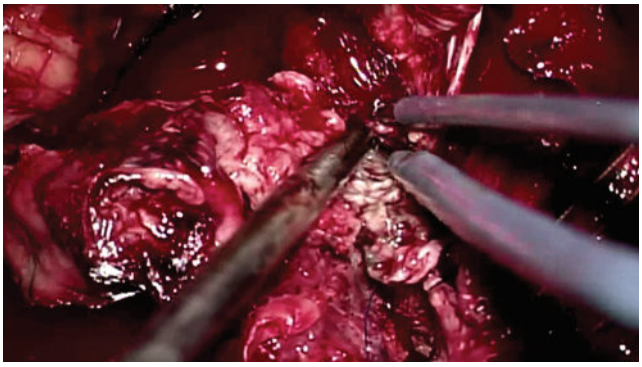


Fig. 28.11 Intraoperative view of tumor resection from within the transverse sinus. The tumor was delivered as the incision line was closed with Prolene suture seen in the bottom aspect of the image.

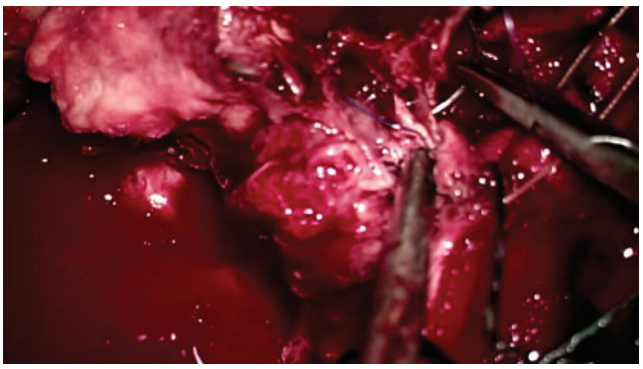


Fig. 28.12 Intraoperative view of sinus closure.

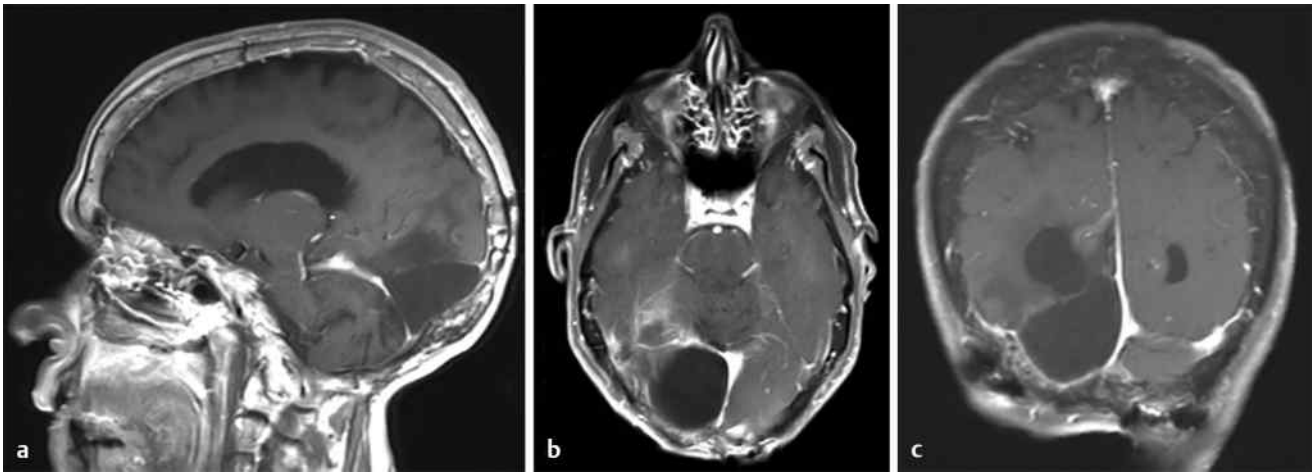


Fig. 28.13 Postoperative **(a)** sagittal, **(b)** axial, and **(c)** coronal gadolinium-enhanced MRI showed gross total resection. Residual tumor was purposefully left on the straight sinus due to its adherence to the deep venous system but is not appreciated on MRI.

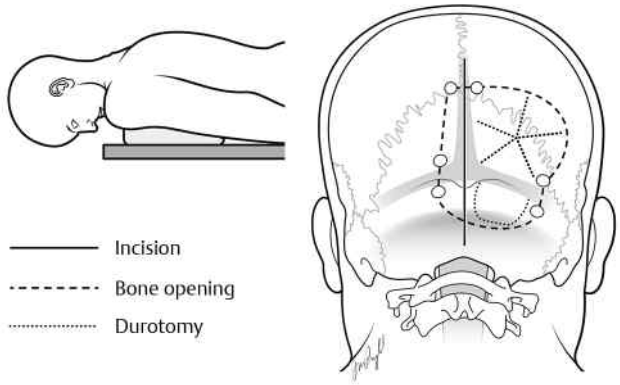
Operative Setup

Position: prone

Incision: straight, midline

Bone Opening: occipital and suboccipital

Durotomies: Stellate over tumor and suboccipital



ultrasound subse quently confirmed sinus patency. Any tumor that infiltrated the tentorium and falx was then resected. A small amount of tumor was found adherent to the straight sinus, which, by Doppler examination, had significant flow. Since there was no collateral system to handle this flow in the straight sinus, the decision was made to leave the small residual behind.

Following resection, thorough hemostasis was performed and the wound was copiously irrigated. The posterior fossa dura was repaired with the aid of an implantable dural substitute. The bone flap was then fixed to the skull using titanium plates and the wound closed.

Surgical Pearls

1. Proper preoperative planning must necessarily include discussion with the anesthesia team regarding the opening of a large venous sinus during surgery. Preparation must be made for monitoring and potential intervention for venous air embolism, as well as for significant blood loss after flow is restored in a previously stenotic or occluded sinus.
2. Ophthalmological examination is helpful to assess for baseline visual function, as a partial hemianopsia may not be apparent to the patient before surgery. For proper management of expectations, patient and family should be counseled on the risk of visual changes or temporary cortical blindness as a result of brain retraction or vascular injury.
3. During the resection, tumor draining veins must be identified and differentiated from veins en passage, so that the latter can be preserved to avoid venous infarction.
4. In patients like ours where there is hydrocephalus, preoperative cerebrospinal fluid shunting is not advisable as surgical debris from the resection often damages the shunt. External ventricle drainage should be used instead, and if the patient is dependent on the external ventricular drain after the resection, placement of a permanent shunt can be performed shortly after the resection.

■ Aftercare

Postoperative CT imaging was obtained to identify any venous infarcts and fortunately, there was none. Gadolinium-enhanced MRI after surgery showed no residual as the tumor left on the straight sinus was not visualized (**Fig. 28.13**). There were no postoperative complications. Histopathological examination showed a WHO grade I meningioma, and the patient continues to be followed with serial MRIs. No adjuvant radiation was recommended unless tumor regrowth was identified. Postoperative ophthalmologic exam showed a partial left homonymous hemianopsia. Her preoperative symptoms of headache and ataxia both resolved.

Perspective

Ilyas M. Eli and William T. Couldwell

■ Introduction

Meningiomas that invade the torcula occur infrequently, and treatment of these rare lesions requires extensive preoperative planning and a multidisciplinary approach. Multimodality imaging is critical to determine the anatomical nuances of the tumor's extensions and its vascular infiltration. Some authors advocate treating torcular meningiomas with a strategy of tumor debulking to reduce mass effect followed by radiosurgery, but long-term data for this paradigm is lacking. Furthermore, for large meningiomas, radiation can sometimes exacerbate the peritumoral edema. Our goal of management of all meningiomas is to achieve a complete or low Simpson grade resection, and to preserve neurological function. The primary determinant for the extent of resection of torcular meningiomas is the relationship between the tumor and the venous sinuses, which, despite high-quality multimodality preoperative imaging data, often requires intraoperative verification.

■ Case Presentation

A 48-year-old man presented with several days of worsening headache and blurred vision. The findings on physical examination were notable for a firm protuberance in the occiput without any skin abnormalities. On neurological examination, he had an inferior field hemianopsia. MRI and MR angiography revealed an 11 × 5-cm mass involving the bilateral parietal and occipital lobes, as well as the SSS, the left proximal transverse sinus, and the torcula (**Fig. 28.14**). Additionally, the mass had transosseous extension into the calvaria and into the adjacent scalp and dermis. DSA revealed absent flow in the SSS, torcula, and proximal transverse sinuses (**Fig. 28.15**). Deep venous outflow passed through an occipital sinus and cortical venous drainage via the vein of Labbé.

Our differential diagnosis based on the MRI included meningioma, hemangiopericytoma, sarcoma, and possible malignancy. In addition to intracranial imaging for characterization of the mass, CT of the chest, abdomen, and pelvis was obtained, which did not show evidence of tumor elsewhere.

■ Anatomical and Therapeutic Considerations

Embolization

DSA and venography serve as important diagnostic tools to visualize the patency of the venous sinuses involved, and to assess the vascular supply of the tumor, which can be embolized preoperatively to reduce vascularity. Furthermore, angiography is helpful in identifying abnormal drainage through venous collaterals. In this patient, the angiogram showed no flow in the SSS, torcula, and left proximal transverse sinuses. Furthermore, the mass was hypervascular, with arterial supply from the meningeal branches of the external carotid artery and posterior inferior cerebellar arteries. Preoperatively, the vascular feeders of the meningioma from the external carotid artery were embolized with coils and polyvinyl alcohol particles.

Perioperative Preparation

The patient must be counseled about the potential complications of surgery related to further venous sinus occlusion. Since the surgery takes place next to large venous channels, air embolism must be considered, and therefore precordial Doppler probe and central venous catheter are used for its detection. The planned position is prone, with the head of the patient maintained below the level of the heart to lower the risk of air embolism. The senior author prefers to initiate aspirin (650 mg the day before surgery) if sinus violation is planned, and this is continued perioperatively.

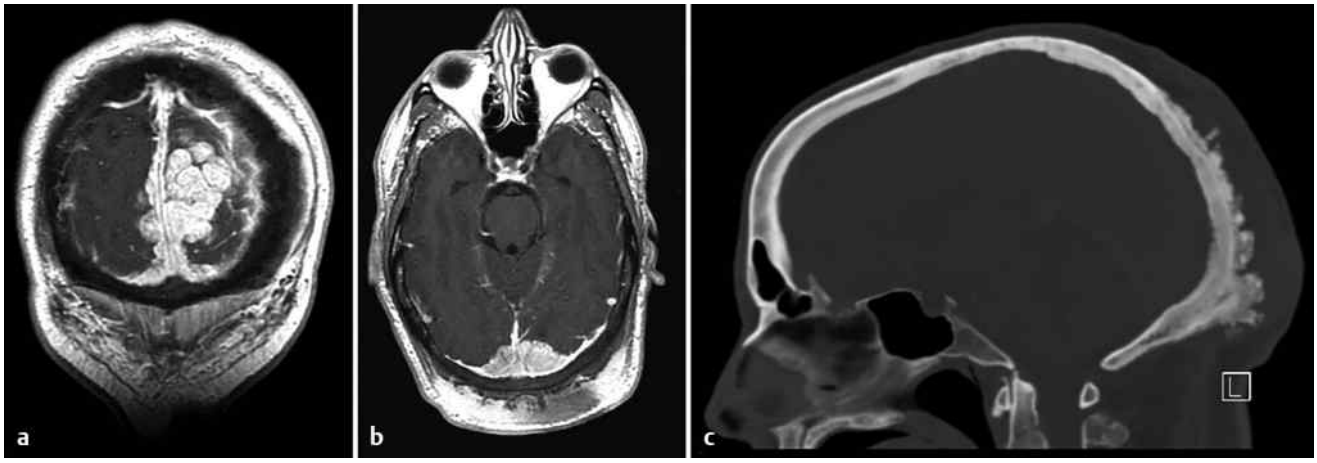


Fig. 28.14 Preoperative images. **(a)** Coronal and **(b)** axial T1-weighted MRI with contrast, demonstrating tumor involving the posterior portion of the superior sagittal sinus and torcula. **(c)** Sagittal CT scan demonstrating hyperostotic tumor infiltration of the occipital bone.

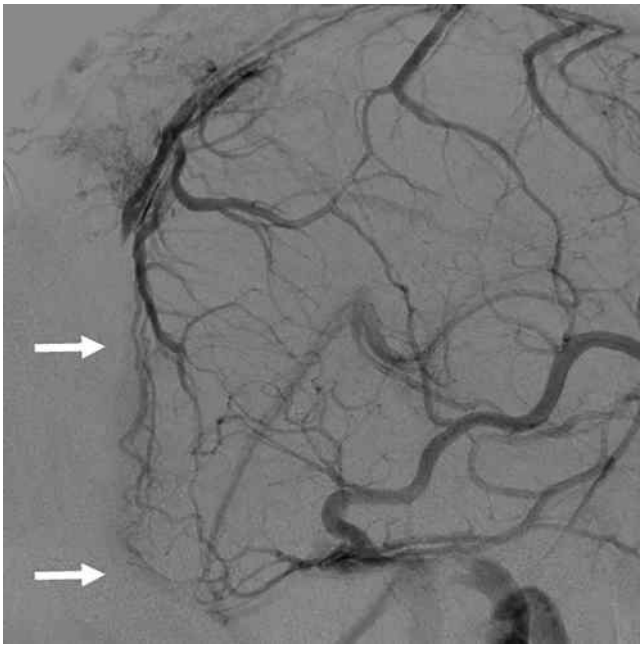


Fig. 28.15 Preoperative angiogram. Digital subtraction angiogram (lateral view) demonstrated occlusion of the posterior SSS, torcula, and proximal left transverse sinus (*white arrows*). Deep venous drainage passed through the occipital sinus connecting the straight sinus to the left jugular vein.

Venous Sinus Management

Some proponents argue for an aggressive or radical approach for complete tumor resection, whereas those on the other end of the spectrum advocate a more conservative approach, with tumor debulking of the extravascular portions and treatment of the residual tumor with adjuvant radiation and/or observation. In cases where an aggressive approach is taken, management of the venous sinus is also controversial. The authors of the preceding section described their technique for removing tumor from a nearly occluded transverse sinus with subsequent sinus reconstruction. It is generally accepted that patients, who have a complete occlusion of the sinus with venous drainage diverted to developed collaterals, can tolerate resection of the involved sinus without the need for sinus reconstruction. The overarching goal is to avoid further venous outflow obstruction (or propagation of intraluminal clot beyond the region of occlusion), which could result in a poor neurological outcome, such as edema, venous infarction, or even death.

In a case where the tumor has already occluded the sinus, verified by venography, our preference is to ligate and resect a completely occluded sinus without reconstruction. Intraoperatively, if there is contradicting but clear evidence of sinus patency, then sinus reconstruction is performed, since collateralization may not be complete. Especially in younger patients, we prefer to be aggressive and obtain a gross total resection. The goal is to reduce the risk of recurrence in patients with a long life expectancy and avoid radiation in this age group. In older patients, debulking the tumor followed by radiation treatment may be a reasonable option.

In patients with tumor infiltrating one wall of the sinus and incomplete sinus occlusion, we prefer resection of the infiltrating tumor followed by sinus reconstruction. However, unlike the authors of the previous section, we repair the sinus defect primarily with a dural graft (autologous dura or fascia preferentially) or using an allogeneic tissue patch. The proximal portion is closed first to allow for back-bleeding during the suturing. We suture the vessel wall using a Prolene 6–0 or 7–0 monofilament suture in a continuous fashion. To prevent sinus thrombosis, 650 mg of aspirin is administered preoperatively or per rectum with intravenous heparin bolus prior to the repair. The patient is also continued on 325 mg of aspirin daily for several weeks after surgery. For management of venous sinus bleeding during the repair, clips may be gently placed across the sinus (to avoid tearing of the sinus or injury to the wall of the sinus), or cottonoids may be used for temporary occlusion to reduce venous bleeding.

Transosseous and Transdermal Infiltration

Understanding the total extent of tumor invasion is important to achieve a full resection. The MRI, CT scan, and clinical findings indicated that our patient had transosseous, as well as transdermal, infiltration of the tumor. A wide excision of the involved portions of the dura, calvaria, and skin was required to prevent recurrence. The dura can be reconstructed using AlloDerm and the removed bone can be replaced with mesh cranioplasty. Large skin defects require the assistance of plastic surgeons for reconstruction of the scalp using a rotational flap or vascularized free flap.

■ Description of the Technique

The patient was placed in the prone position using a Mayfield three-pin head holder. Image-guidance was utilized, and a wide scalp excision and craniectomy were performed. The involved skin was removed with wide margins. The underlying bone was hyperostotic with areas of soft tumor infiltration. Using neuronavigation, the SSS, torcula, and transverse sinus were outlined, and burr holes were placed along the sinuses and periphery to encompass portions of both the supra- and infratentorial compartments. After the craniectomy exposing the tumor and the region of the torcula, the dura was opened over the bilateral parietal and occipital lobes. Arachnoid violation by the tumor was noted throughout, with parasitized dilated cortical arterial feeding vessels. The tumor was resected using standard microsurgical technique while preserving the surrounding veins (**Fig. 28.16**).

The proximal normal SSS was ligated using a 3–0 Vicryl suture. The sinus and falx were incised down to the straight sinus, which had no evidence of tumor invasion and was preserved. Resection proceeded into the torcula and left transverse sinus, which was occluded by extensive tumor invasion. The involved walls were ligated and incised along with the tentorium and tumor. The right transverse sinus was not invaded and thus preserved. Its connection to the torcula was closed primarily using 6–0 Prolene sutures. Duraplasty was performed with AlloDerm, and then a titanium mesh cranioplasty was placed. The plastic surgery team repositioned the patient in the lateral decubitus position to perform a left latissimus dorsi free flap. The free flap blood supply was provided via

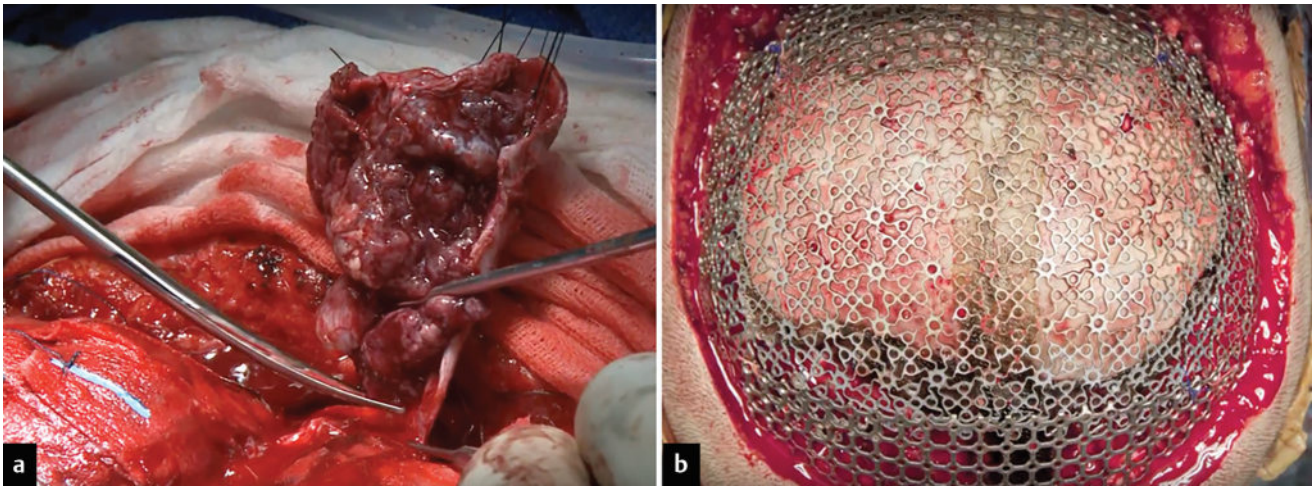


Fig. 28.16 Intraoperative views. (a) Resection of the tumor and dural attachment. **(b)** AlloDerm duraplasty and titanium mesh cranioplasty.

anastomosis of the left superficial temporal artery to the thoracodorsal artery using an 8–0 nylon suture. The thoracodorsal vein was anastomosed to the superficial temporal vein using a 2-mm venous coupler. A halo vest was applied to protect the flap from pressure necrosis.

■ Aftercare

The patient was transferred to the neurocritical care unit. He experienced decreased visual acuity, which improved during his inpatient rehabilitation course. The halo vest was continued for 6 weeks. Final pathological analysis indicated the lesion was a WHO grade I meningioma.

■ Commentary

Operative management of torcular meningiomas is technically challenging. The optimal management is to achieve a complete or low Simpson grade resection with preservation of the patient's neurological function, if feasible. Where the edge of this feasibility lies and what Simpson grade is low "enough" are topics debated in the literature to this day, as proposed treatment paradigms vary widely from tumor debulking leaving the torcula and involved sinus untouched, to radical tumor resection and sinus excision, with or without sinus reconstruction. Our case illustrates the senior author's surgical approach and preference for an aggressive resection of torcular meningiomas, tempered with the realization that this may not be possible without venous consequences. Regarding the case presented by the authors of the preceding section, we would have managed that patient with near-occlusion of the right transverse sinus in much the same way, and despite the patient's age, would remove the tumor from the sinus instead of leaving that for radiation. However, our preference is to repair the sinus with a dural graft over primary closure.

In approaching torcular meningiomas, extensive preoperative preparation is required to evaluate the venous drainage pattern, which in turn determines optimal handling of the tumor and sinus(es). Intraoperatively, even conservative and careful tumor debulking can lead to venous sinus injury. Because of this and regardless of the preoperative surgical plan, surgeons must be prepared to handle, repair, or resect the involved venous sinus. Preparation must necessarily include having on hand a full set of vascular instruments, sutures, and sinus reconstruction graft material (autograft or other). Sinus bleeding should be controlled by gently placing vascular clips or cottonoids for temporary occlusion during tumor removal and reconstruction. Coordination with the anesthesia team is paramount, so that they too can be prepared, whether it is to help manage venous air embolism (bed control, central line, Doppler echocardiography), to have blood readily available for expeditious transfusion, or to deliver rectal aspirin or intravenous heparin.

Finally, successful management of torcular meningiomas is achieved by multidisciplinary discussion of the patient in tumor board and utilizing a team-based approach. The stakes are high when the operation takes place around large venous channels and the torcula. Resecting what appeared to be a completely occluded sinus may, on rare occasion, result in venous infarction, as bridging veins draining into the tentorium are interrupted. There are a thousand ways to fail with these tumors. As such, a team-based approach, with thoughtful discussion among neurosurgeons, endovascular radiologists, and radiation oncologists, is likely the patient's best protection against management risks, and the path to optimize clinical outcome.

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Part IX

Tumors of the Ventricles

29 Lateral Ventricle (Monro) *365*

30 Lateral Ventricle (Atrium) *376*

31 Third Ventricle *390*

32 Fourth Ventricle *402*

29 Lateral Ventricle (Monro)

R. Tushar Jha and Walter C. Jean

Keywords: fornix, tela choroidea, colloid cyst, arteriovenous malformation, aneurysm, adenoma, endoscope

■ Case Presentation

A 48-year-old woman with a past medical history of migraines and fibromyalgia presented to the neurosurgery clinic with the chief complaint of headaches and emesis. Her first episode of headache was about 5 months prior, and it went away like a "normal" headache. One day in the following month, she recalled getting a headache, and she vomited when the headache was at its peak. Since then, she has had daily morning headaches, which she attributed to "allergies." On direct questioning, she denied any "thunderclap" onset of any of her headaches. She said they were all gradual in onset and progressed before they went away again. Because of this history, her physician ordered cranial imaging and thereafter referred her to our unit.

On physical examination, she was neurologically intact. Computed tomography (CT) and magnetic resonance imaging (MRI) scans of the brain revealed a lesion of the foramen of Monro and flow voids in the right parietal lobe (**Fig. 29.1**). There were no findings of an acute ischemic or hemorrhagic stroke. The ventricles were normal in size.

Questions

1. What are the differential diagnoses of the MRI findings and are there any further studies that should be performed?
2. Which lesion do you think is more likely to be culpable for her headaches?

■ Diagnosis and Assessment

At time of presentation, the CT and MRI showed two distinct lesions—a well-demarcated, well-circumscribed mass in the foramen of Monro region and a vascular lesion of the right parietal lobe. The foramen of Monro lesion measured 9.3 mm in diameter on CT scan (**Fig. 29.1**). MRI showed that this lesion was hyperdense on FLAIR, but did not enhance with contrast. Lesions of the foramen of Monro can be vastly different between pediatric and adult patients. The most common lesion around the foramen of Monro in adults is a colloid cyst. These represent 15 to 20% of intraventricular masses and are located in the foramen of Monro in 99% of cases. The majority of cases are identified between 30 and 40 years of age. Typically, colloid cysts appear hyperdense on CT and are attached to the anterior roof of the third ventricle. On MRI, these lesions are usually hyperintense on T1 and isointense on T2 due to its thick "motor oil" fluid consistency. Colloid cysts typically do not enhance with contrast, but may occasionally display rim enhancement.

Rare lesions of the foramen of Monro include meningioma, subependymal giant cell astrocytoma, juvenile pilocytic astrocytoma, choroid plexus papilloma, and central neurocytoma. Since the majority of these enhance with gadolinium administration on MRI, they can be ruled out. Central neurocytomas are low grade lesions and they cannot be completely ruled out, as the enhancement pattern is less consistent. However, the majority of neurocytomas display mild to moderate heterogeneous enhancement, and they often contain numerous cystic areas that have a bubbly appearance and can completely attenuate on FLAIR MRI. All said, given the lesion's location and imaging characteristics in this patient, it was most likely a colloid cyst.

The MRI flow voids and time of flight MRI findings in the right parietal lobe of our patient are consistent with an arteriovenous malformation (AVM) (**Fig. 29.1b,c**). For this reason, an angiogram was obtained and this revealed a right parietal AVM with a 3-cm nidus and feeders from the angular branch of the middle cerebral artery (MCA), parietal occipital branch of the MCA, and pericallosal branch of the right anterior cerebral artery. There was also a superiorly projecting feeding artery, flow-related, prenidal aneurysm coming off the M1 MCA segment, just prior to the bifurcation (**Fig. 29.1c**). The dome and neck of this aneurysm measured 6 and 3 mm, respectively. This AVM displayed superficial venous drainage into the superior sagittal sinus (SSS). On the Spetzler–Martin (SM) AVM grading system, this AVM was classified as grade II.

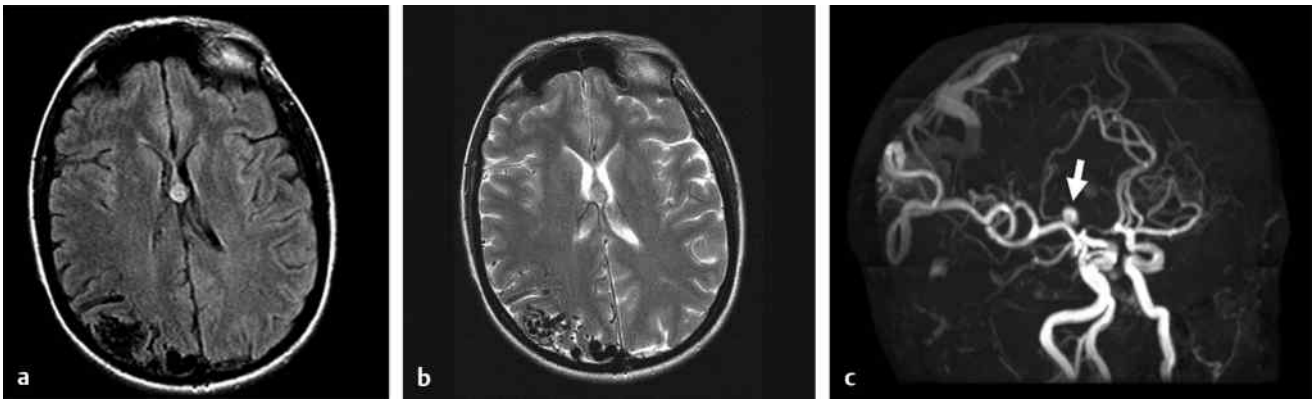


Fig. 29.1 Preoperative images. (a) Axial FLAIR image showing a hyperintense lesion at the foramen of Monro. Note the abnormality in the right parietal lobe; **(b)** T2 image not only redemonstrated the ventricular lesion, but also showed the flow voids in the right parietal lobe consistent with an arteriovenous malformation (AVM); **(c)** MRA demonstrated the AVM and *white arrow* highlights the flow-related, perinidal aneurysm.

Questions

1. Does this patient's colloid cyst require intervention? Or is "watchful waiting" better for her?
2. If surgery is indicated for the cyst, what approach is best given the normal-sized ventricles?
3. What is the most dangerous aspect of the patient's AVM?
4. Does the AVM require intervention? What about radiosurgery?

■ Anatomical and Therapeutic Considerations

The Treatment Dilemma

Our patient had two distinct and separate lesions—a foramen of Monro colloid cyst and a right parietal SM grade II AVM with an associated flow-related, prenidal aneurysm. Ultimately, there are several questions that arise from the unique "double trouble" aspect of this case:

1. Which lesion, if any, was responsible for her symptoms? Does it require intervention?
2. Does the other problem need treatment? If so, when?
3. If the colloid cyst needs to be treated first, how would the AVM/aneurysm impact on that treatment? Does it affect the approach?
4. Conversely, if the AVM/aneurysm needs to be treated first, how would the colloid cyst impact that treatment? Does it affect the approach or modality?

The Colloid Cyst

Our patient's colloid cyst was discovered on workup for headaches, nausea, and emesis. Nonetheless, she was neurologically intact and radiographic scans did not show evidence of hydrocephalus. While colloid cysts are histologically benign, they are located at the foramen of Monro where they can have a "ball–valve" effect and cause acute obstructive hydrocephalus, rapid neurological deterioration, or even sudden death. The risk of rapid neurological deterioration varies widely from 3 to 35% and the estimated risk of sudden death ranges from 5 to 10%.

On the other hand, surgical resection of this deep-seated lesion near the fornix has a major complication risk of 15%. Therefore, it is especially important when managing colloid cysts to weigh the risks and benefits of expectant management against those of surgical resection. But thanks to the work of Beaumont et al, there exists a tool to gauge the danger this colloid cyst posed to the patient and determine if intervention was justified or necessary. The five-point Colloid Cyst Risk Score (CCRS) stratifies the risk for sudden, catastrophic hydrocephalus, and helped us to formulate a plan for the patient (**Table 29.1**).

This scale takes into account the five most significant risk factors for symptomatic colloid cysts and assigns a point for each risk factor that is present. These risk factors include age less than 65 years, presence of headache, cyst's axial diameter of greater or equal to 7 mm, presence of FLAIR hyperintensity, and cyst located in an anatomical risk zone. The third ventricle risk zones are defined by dividing the structure into three regions based on sagittal plane T1 MRI. Zone I extends from the lamina terminalis to a vertical line drawn from the mammillary body tangential to the massa intermedia and includes the most common location for these cysts—the foramen of Monro; zone II extends from the posterior boundary to zone I to the rostral apex of the midbrain tegmentum at the inlet of the cerebral aqueduct; zone III extends from the posterior boundary of zone II to the posterior commissure.

Table 29.1 The colloid cyst risk scale (CCRS)

Criterion	Points
Age < 65 years	
• Yes	1
• No	0
Headache	
• Yes	1
• No	0
Axial diameter ≥ 7 mm	
• Yes	1
• No	0
FLAIR hyperintensity	
• Yes	1
• No	0
Risk zone	
• Yes	1
• No	0

Note: CCRS ≤ 2 is low risk subgroup. CCRS ≥ 4 is high risk subgroup. CCRS of 3 is the intermediate risk subgroup.

Based on this scale, our patient received points for age less than 65 years, presenting with a headache, axial diameter of 9.3 mm (≥ 7 mm), cyst FLAIR hyperintensity, and cyst location in a risk zone (zone I). This characterized our patient's colloid cyst as a CCRS 5 and objectively identified it as a symptomatic lesion with a high risk for obstructive hydrocephalus and neurological deterioration. Expectant management for this lesion would not be appropriate and surgical removal of the cyst was appropriate and necessary.

Options for Approach

The two general approaches to the foramen of Monro are the interhemispheric transcallosal and the frontal transcortical, transventricular approaches (**Fig. 29.2**). The interhemispheric transcallosal approach is an elegant approach exploiting the natural dissection plane between the falx cerebri and the interhemispheric fissure (**Fig. 29.3**). Since it is performed near the midline, the approach offers an excellent view of the foramen of Monro, often well into the contralateral side. The only brain tissue that is disrupted by the approach is a small stretch of the anterior body of the corpus callosum, and disconnection syndrome is exceedingly rare. One disadvantage of the approach is potential disruption of draining veins between the frontal lobe and SSS, which, on rare occasions, can lead to venous infarction. The pericallosal arteries are also in harm's way during this approach, and disruption and narrowing of one or both arteries during surgery can sometimes lead to significant leg weakness in the patient.

Contrastingly, neither the sagittal draining veins nor the pericallosal arteries are in danger in the frontal transcortical, transventricular approach which can be performed either by microsurgical or endoscopic techniques. The main

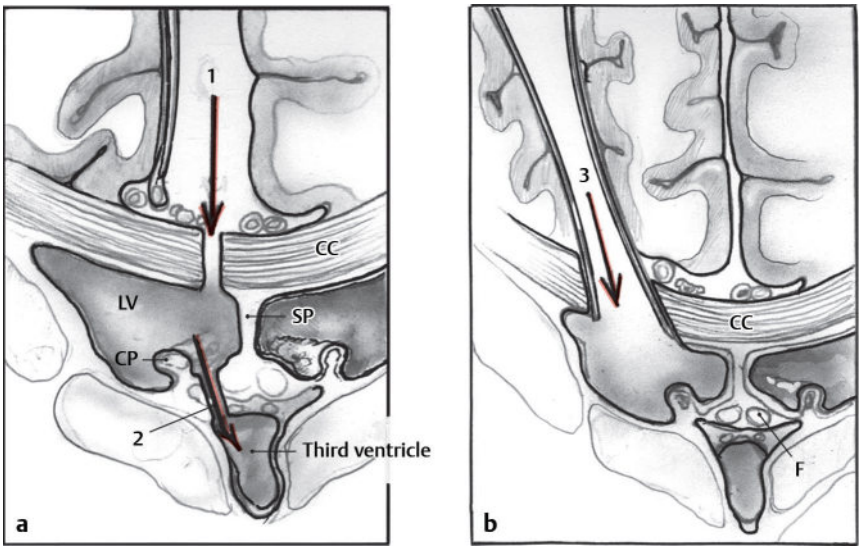


Fig. 29.2 Microsurgical approaches to the foramen of Monro. Coronal view illustrating the routes of entry into the region of the foramen of Monro and anterior third ventricle. The right lateral ventricle is cut at the level of the foramen of Monro and the left lateral ventricle at the massa intermedia. **(a)** 1, transcallosal route; 2, transforaminal route. **(b)** 3, transcortical route. CC, corpus callosum; CP, choroid plexus; F, fornix; LV, left ventricle; SP, septum pellucidum. (Reproduced from Nader R et al, eds. Neurosurgery Tricks of the Trade. Thieme; 2014.)

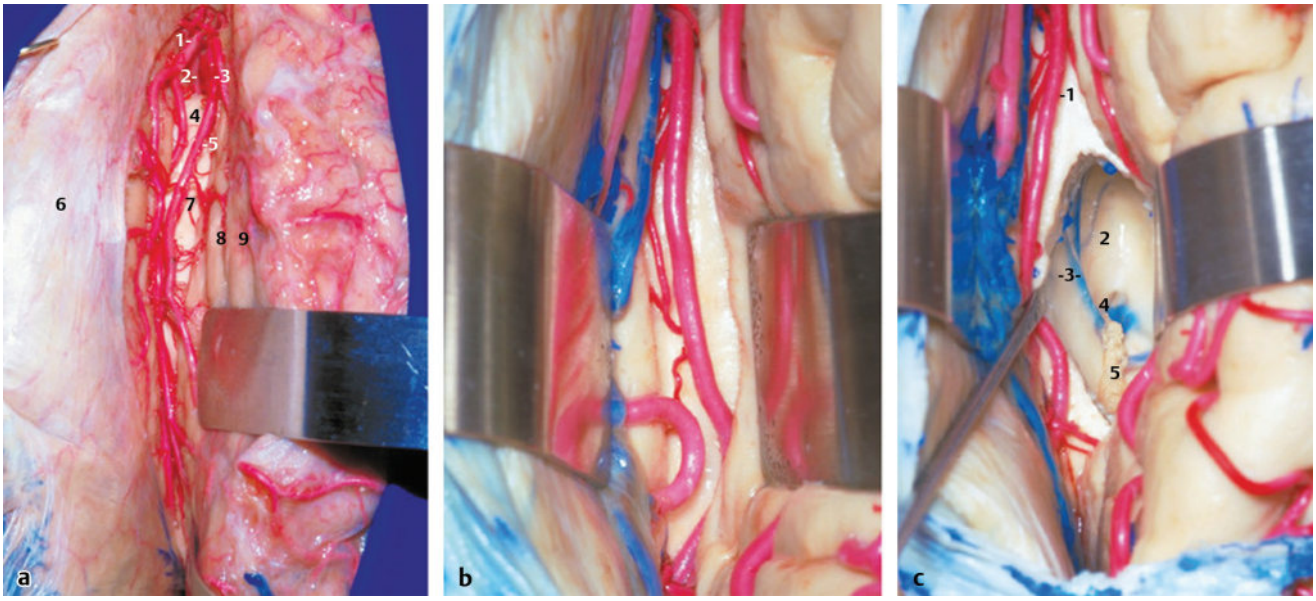


Fig. 29.3 Anatomical dissection demonstrating the interhemispheric transcallosal approach. (a) 1, left pericallosal artery (A3 segment); 2, left pericallosal artery (A2 segment); 3, right pericallosal artery; 4, genu of the corpus callosum; 5, right pericallosal artery (A4 segment); 6, falx; 7, body of the corpus callosum; 8, cingulate gyrus and sulcus; 9, medial frontal gyrus. **(b)** Anatomical dissection of the interhemispheric fissure. A magnified view of the body of the corpus callosum after separating both cingulate gyri. **(c)** View into the right lateral ventricle after callosotomy. The frontal horn and the body of the lateral ventricle are exposed. 1, pericallosal artery; 2, rostrum of the corpus callosum; 3, septum pellucidum and anterior septal vein; 4, foramen of Monro; 5, choroid plexus of the body of the lateral ventricle. (Reproduced from Nader R et al, eds. Neurosurgery Tricks of the Trade. Thieme; 2014.)

weakness of this approach is, since it is nearly universally performed through the right side, difficulty in visualizing the left frontal horn, and this may be particularly problematic for lesions with significant bilateral components. Although there is conflicting data whether this approach is associated with increased risk for postoperative seizures, it does traverse significant amount of brain tissue to reach the lateral ventricle. As such, the frontal transcortical, transventricular approach is usually reserved for patients with significant ventriculomegaly, in whom the dilatation of the frontal horn and foramen of Monro itself mitigate the effects of these disadvantages. The endoscopic variant of this technique minimizes tissue damage, but the visualization of the contralateral side remains limited, and through the endoscope, bleeding on that side might be difficult to control.

For the above reasons, the interhemispheric transcallosal transforaminal approach was chosen to remove the colloid cyst, but the concern remained whether the AVM should be treated first. Even if the answer is "no," would the AVM and the associated prenidal aneurysm pose a danger to the patient with the hemodynamic and intracranial pressure fluctuations during colloid cyst surgery?

The Arteriovenous Malformation

The patient's grade II AVM had a flow-related, prenideal aneurysm coming off the right M1 artery, and it was this feature of the AVM that posed the most danger to the patient. It also had dilated cortical venous drainage to the SSS. The average annual rupture rate of AVMs is estimated to be 2 to 4%. However, the prenidal

aneurysm in the case of our patient's AVM increases the annual rupture rate to about 7%. The lifetime AVM rupture risk can be calculated using the formula [1– (risk of no ruptures)^n]. Assuming a life expectancy of 78 years for our patient, and taking into account her age of 48 years at presentation, and the AVM annual rupture risk rate of 7%, this patient's AVM lifetime rupture risk is [1– $(0.93)^{30}$] or 88.67%. This is a rupture risk that warrants treatment. But when should it be treated and with which modality?

Options for Approach

The component of the AVM most likely to bleed was the prenidal aneurysm. Rupture of this aneurysm would cause subarachnoid hemorrhage, which has a 30% risk of morbidity regardless of posthemorrhage treatment. The digital subtraction angiography revealed a 6 mm aneurysm dome and a 3 mm aneurysm neck. The dome to neck ratio of 2 to 1 and a location proximal to the MCA bifurcation were both features of this prenidal aneurysm that made endovascular coiling the preferred treatment modality.

The remainder of the AVM can be treated using microsurgical resection, endovascular coiling, radiosurgery, or a combination of these. Radiosurgery treatment for AVM has the benefit of being noninvasive, but it can take up to 3 years to achieve complete thrombosis, during which time the patient remains at risk for hemorrhage. For our patient, this treatment might be considered after her colloid cyst has been eliminated, but in the short term, it would do nothing to protect her before or during surgery for her colloid cyst. Given the grade of the patient's AVM, the risks of microsurgical resection were acceptably low. Furthermore, preoperative embolization may reduce the risks even further, and therefore a combination treatment of staged embolization, followed by microsurgical resection was the most sensible way to eliminate the lesion.

Timing and Sequence of Treatment

Based on the timing, pattern and other characteristics of the patient's headaches, they were most likely linked to the colloid cyst. She has never had a "thunderclap" headache, describable as the "worst in her life," and the headaches occurring every morning were quite suggestive of positional obstruction of the foramen of Monro by the cyst during recumbence. In addition, the cyst was a CCRS 5 lesion, and therefore a symptomatic lesion placing our patient at high risk for clinical deterioration. All this suggested that the colloid cyst was culpable for the patient's acute symptoms, and logically, it should be treated first.

However, surgery on the colloid cyst would involve predictable release of cerebrospinal fluid, and unpredictable hemodynamic and intracranial pressure shifts during the perioperative period. Either of these can alter the transmural pressure across the wall of the prenidal aneurysm, causing this most precarious part of the AVM to rupture. In this regard, even though her headaches were unrelated, the prenidal aneurysm might in fact pose the most danger to the patient. With this consideration, a stratification of the most-threatening to the least-threatening lesions was beginning to emerge: prenidal aneurysm, colloid cyst, AVM. This was, logically, the sequence in which these lesions should be eliminated.

In summary, our patient's lesions were treated in the following sequence:

- Hospital stay 1:
  - 1st: Endovascular coiling of right M1 prenidal aneurysm
  - 2nd: Microsurgical resection of colloid cyst
- Hospital stay 2:
  - Stage I endovascular embolization
- Hospital stay 3:
  - Stage II endovascular embolization
- Hospital stay 4:
  - Microsurgical resection of AVM

Questions

1. How would you handle the bridging veins to the SSS?
2. What is the best way to identify the corpus callosum?
3. Where would you place the craniotomy for the transcallosal approach?

Description of the Technique

Following the plan already laid out, the patient's prenidal aneurysm was obliterated via endovascular techniques without neurological sequela. The next day, she underwent the surgery for the colloid cyst (see **The Three Approach Elements**). After general endotracheal anesthesia was established, the patient was placed in supine position. Her head was fixed in the neutral position. The top of the head was prepped and draped in usual sterile fashion with hair-sparing technique. A linear incision was made parallel to the coronal suture, extending from the left midpupillary line ending 4 to 5 cm above the right tragus. The sagittal and coronal sutures were visualized. The craniotomy was made on the right starting with three burr holes on the midline, one at the coronal suture, and the others anterior and posterior to it. These holes were used to carefully dissect the SSS away from the cranium. A high-speed drill was then utilized to fashion a craniotomy 5 cm long and 3 cm wide, two-thirds in front and one-third behind the coronal suture. In the midline, the bone opening ended just left of the SSS.

The Three Approach Elements

Corridor: interhemispheric

Craniotomy: 5cm × 3cm straddling coronal suture

Modifier: corpus callosotomy

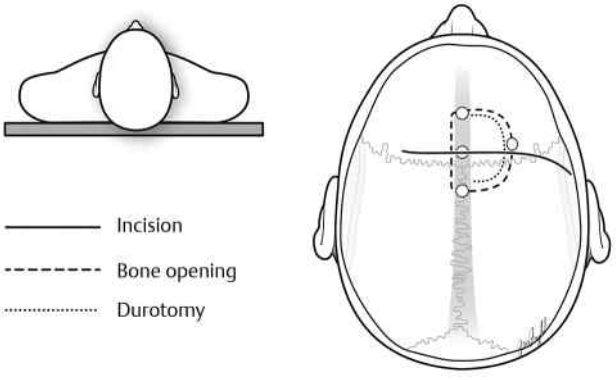
Operative Setup

Position: supine, head neutral

Incision: parallel to coronal suture

Bone opening: right midline, straddling coronal suture

Durotomy: "U-shaped" based on midline/SSS



The dura was then opened in a U-shaped fashion based along the midline. A small bridging vein had to be coagulated, and incised to release the dural fold. Sacrifice of cortical veins draining into the SSS should be avoided, and luckily, none were encountered in this patient (see **Operative Setup**).

At this point, the falx cerebri and interhemispheric fissure were identified (**Fig. 29.4a**). The right frontal lobe was gently mobilized away from midline and a retractor blade was placed on the falx itself to move it slightly left. The adhesions on the cingulate gyrus were dissected (**Fig. 29.5b**), exposing the pericallosal arteries which were carefully padded away (**Fig. 29.5c**). The corpus callosum was then visualized blocking the end of the interhemispheric fissure, a structure distinctly white (**Fig. 29.5d**). A longitudinal 2 cm incision was then made in the corpus callosum to gain access into the right lateral ventricle. Identifying the choroid plexus and the septal vein medially and the right thalamostriate vein laterally confirmed entry into the right lateral ventricle (**Fig. 29.5**).

The foramen of Monro was visualized by following the choroid plexus, and the colloid cyst was thusly found (**Fig. 29.6**). The material inside the mass was semisolid, and after removing some of it, the cyst was swung into the ipsilateral ventricle. Its return path back into the foramen of Monro was blocked by placing a cottonoid there. From here, the cyst was detached from its vascular pedal on the backside (toward the left frontal horn) and removed totally. Meticulous hemostasis

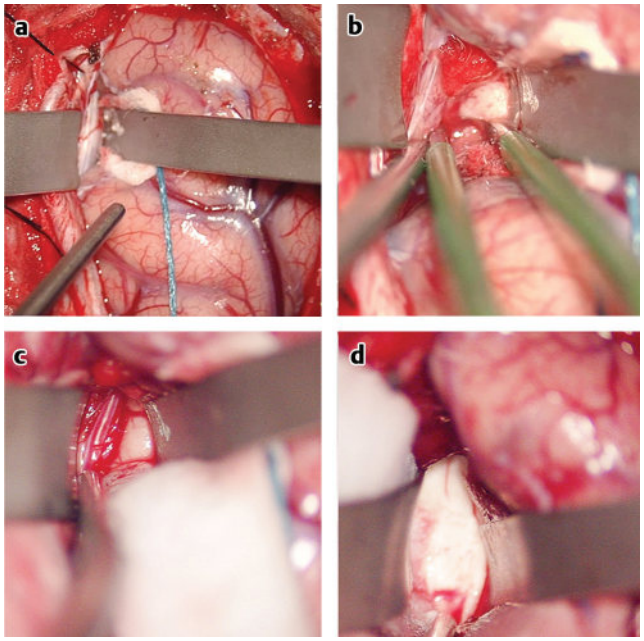


Fig. 29.4 Intraoperative images. (a) Identification of the falx and interhemispheric fissure; **(b)** the cingulate sulcus was dissected to reach the corpus callosum; **(c)** the pericallosal arteries were identified; **(d)** the corpus callosum appeared in its usual stark white color.

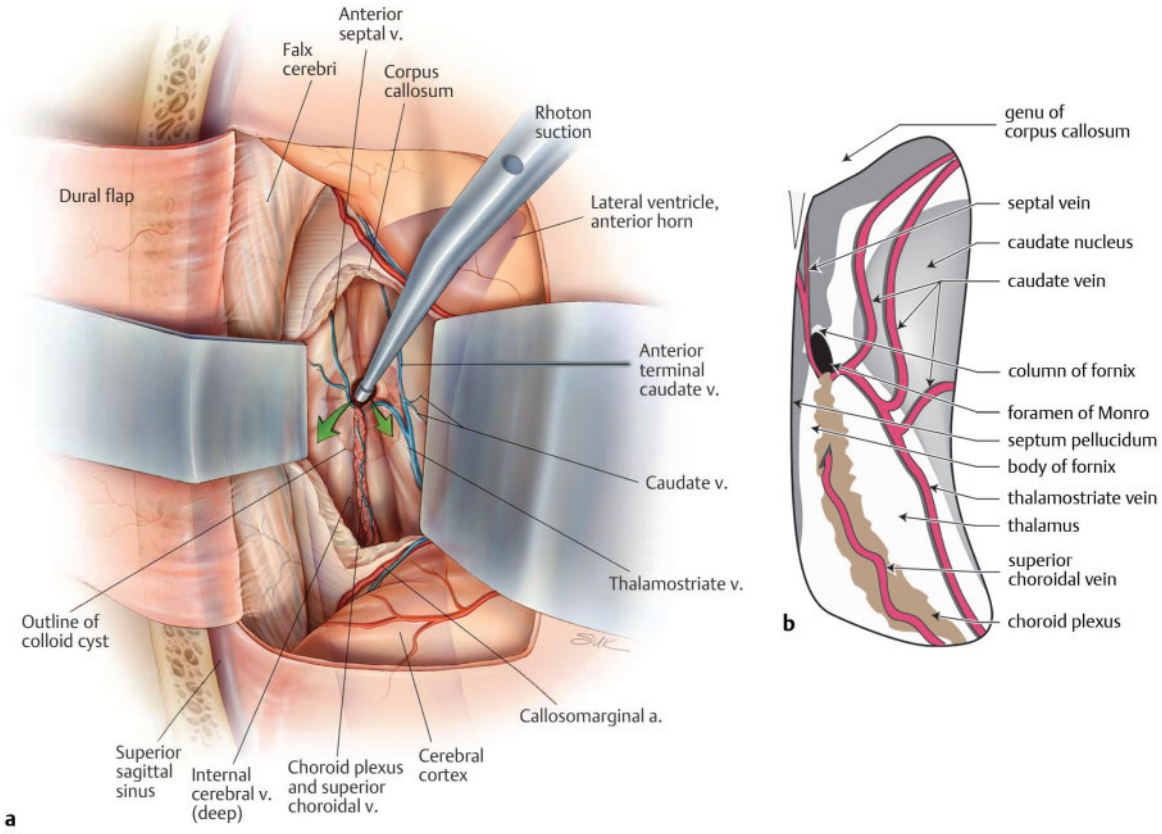


Fig. 29.5 Illustration of the transcallosal, transforaminal approach via the right lateral ventricle. (a) The divided corpus callosum is shown. The choroid plexus is a landmark to the foramen of Monro. The suction is within the foramen of Monro. Tumors can be removed from the third ventricle through an exposed foramen of Monro via a transforaminal approach. (Reproduced from Badie B, ed. Neurosurgical Operative Atlas. Neuro-Oncology. 2nd ed. Thieme; 2006.) **(b)** Schematic diagram depicting the structural relationships within the right lateral ventricle. The foramen of Monro can always be found by following the choroid plexus anteriorly. (Reproduced from Greenberg M, ed. Handbook of Neurosurgery. 8th ed. Thieme; 2016.)

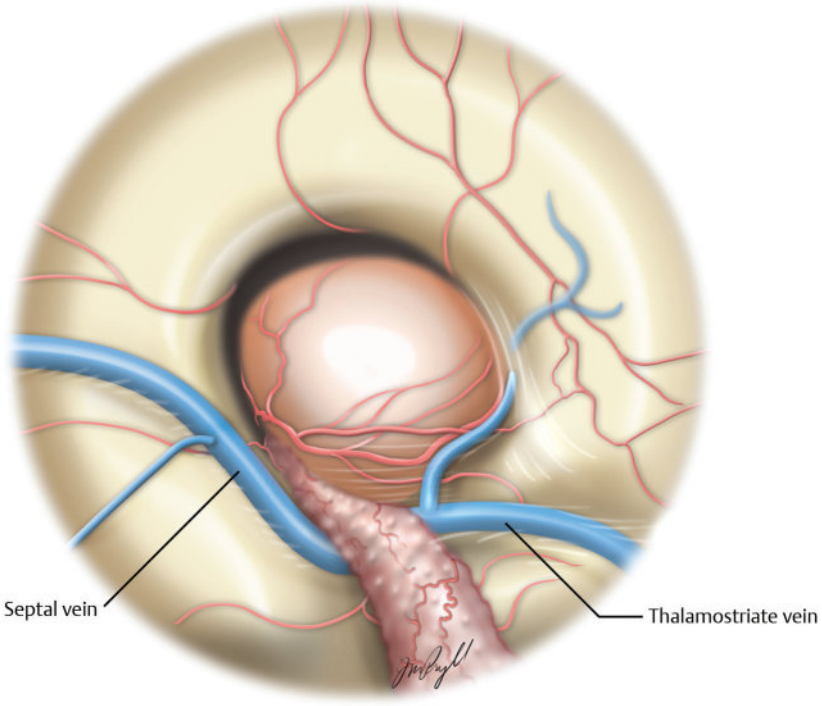


Fig. 29.6 Foramen of Monro. The colloid cyst was seen occupying the foramen of Monro, nearing filling it, without significant expansion of the foramen.

was critical at this point, especially with venous bleeding from the foramen itself. Care was taken not to harm the ipsilateral fornix or thalamostriate vein during hemostasis. Under direct vision and microscopic guidance, a ventricular catheter was placed at the foramen of Monro and secured to the skin. The dura was approximated together with sutures. After the bone flap was replaced the skin was closed in a standard multilayer fashion.

Surgical Pearls

1. In recent years, the senior surgeon has evolved to using the lateral position more often with this procedure, putting the right side down. Gravity-assisted "retraction" naturally mobilizes the frontal lobe away from the interhemispheric fissure, and in addition, makes it less likely for the colloid cyst to fall back into the foramen of Monro or contralateral side.
2. There are a series of anatomical landmarks to keep in mind as one travels down the interhemispheric fissure: first callosomarginal artery, second cingulate gyrus, third pericallosal artery, and fourth corpus callosum. However, the arteries are not always seen and are, therefore, somewhat unreliable as landmarks. One is less likely to confuse the cingulate gyrus from the corpus callosum by remembering that the latter is unmistakably white, while the former is grey.
3. It is the senior surgeon's habit to place three burr holes over the SSS with a round burr on the drill and never the perforator. The middle of the three burr holes is used simply for stripping the SSS, whereas the other two are used for the craniotome during creation of the bone flap. Cutting away from the SSS lessens the risk of injury to the SSS.
4. Sacrifice of any draining veins connecting the frontal lobe and SSS should be avoided. In fact, the length of the craniotomy (5 cm) makes it more likely to find an area devoid of these veins in order to enter the interhemispheric fissure.
5. Since the callostomy is done near the midline, sometimes one lands in the left lateral ventricle instead of the right. For small lesions at the foramen of Monro, this is of little concern. Again, by following the choroid plexus, one finds the foramen regardless of the laterality of the ventricle.

■ Aftercare

After surgery, the patient was neurologically intact from both the aneurysm coiling and the ventricular surgery. The external ventricular drain (EVD) was left in place for 5 days to monitor intracranial pressure (ICP), but especially to drain any residual intraventricular debris. Once the EVD was removed, she was soon discharged from the hospital. Six weeks later, she underwent stage I embolization of the AVM. Approximately 50% of the AVM nidus was obliterated, and the patient was observed overnight. She returned 5 weeks later for a successful stage II embolization and was observed overnight again. Finally, 3 months later, she underwent microsurgical resection of the AVM. She remained neurologically intact after the AVM resection and was eventually discharged to a rehabilitation facility. The two lesions were confirmed on histopathology to be a colloid cyst and AVM and her preoperative headaches had disappeared after all the treatments were completed. At the 2-year mark, she had an angiogram that revealed no vascular abnormalities.

Ten years since her colloid cyst surgery, the patient remains well and fully employed in her thriving career. Her husband complains about her short-term memory issues, so common in patients who have had surgery near the fornix. But otherwise, she is well and her MRI shows no evidence of recurrence of any of her lesions (**Fig. 29.7**).

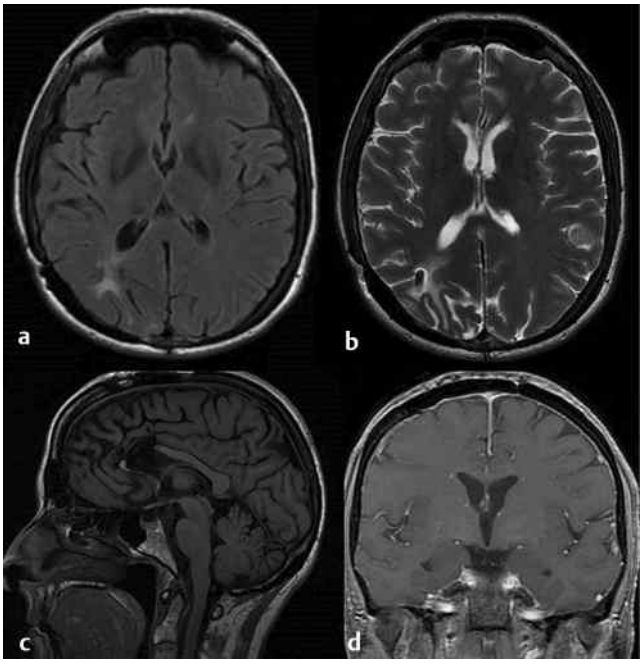


Fig. 29.7 Postoperative images 10 years after surgery. (a) FLAIR axial image showing colloid cyst was removed completely; **(b)** T2 axial image showing arteriovenous malformation had been successfully eliminated as well; **(c)** sagittal image showing 2-cm callosotomy, and **(d)** coronal image showing a "clean" foramen of Monro.

■ Possible Complications and Associated Management

The frontal interhemispheric transcallosal approach involves working around the SSS, which must be handled with care to avoid massive blood loss. Sacrificing bridging veins between the frontal lobe and SSS should be avoided. It is *theoretically* safe to occlude the SSS itself in this anterior location, but on rare occasions, even eliminating a single bridging vein can lead to venous infarction with resultant seizures, motor deficits, or much worse. One of the major risks of the approach is injury to the pericallosal arteries, which can result in contralateral lower extremity weakness. Working around the fornix, there is always the risk for postoperative memory and cognitive problems. With small colloid cysts, there is fortunately rare, and even if it does occur, the disruption is mild as with this patient. It is the senior surgeon's custom to leave the external ventricular drain for 3 to 5 days in order to remove surgical debris from the cerebrospinal fluid and ventricular system. Whether this in fact translates into a lower rate of shunt dependence remains to be proven scientifically.

■ Conclusion

This case highlights the complex decision-making process when multiple lesions are involved. Often, the decisions must be made with imperfect data and in this case, we never knew with absolute certainty that her symptoms were from the colloid cyst. We also had to make an "educated guess" that her AVM would not cause her harm during and immediately after her colloid cyst surgery. Without any evidence of subarachnoid hemorrhage, we decided to eliminate her prenidal aneurysm first, since the coiling was relatively safe, and in our assessment, successful coiling would increase the safety of her colloid cyst operation. The decision-making process for picking the best surgical approach, pervasive in all the chapters in this book, was relatively simply compared to the choice of treatment sequence in this patient.

Perspective

J. André Grotenhuis

■ Introduction

Colloid cysts are benign tumors which almost invariably arise from the tela choroidea of the anterior part of the roof of the third ventricle. They comprise between 0.5 to 2% of all intracranial tumors and 15 to 20% of all intraventricular tumors. Symptomatic colloid cysts often present with the classic symptoms of paroxysmal headache associated with changing head position due to intermittent obstructive hydrocephalus, and with these symptoms, treatment is usually required. As described above, the microsurgical transcortical-transventricular and interhemispheric-transcallosal approaches have been developed as the gold standard to remove colloid cysts at the foramen of Monro. However, over the last two decades, endoscopic removal has emerged as an alternative to craniotomy, and considerable debate exists regarding the overall advantages and disadvantages of open versus endoscopic approaches.

In the previous section of this chapter, the authors presented a patient with a colloid cyst and an AVM with a prenidal aneurysm. They discussed the rationale for the treatment strategy and utilized the transcallosal approach to remove the colloid cyst. Here, we will discuss a similarly complex patient, and highlight the differences in the approach.

■ Case Presentation

This 69-year-old man had an unremarkable medical history except for hypothyroidism for 6 years. But, for the last 2 years, he has experienced progressive gait difficulties with a shuffling walking pattern and short stride length. These symptoms were associated with a decline in short-term memory and urinary urgency. His neurological examination revealed a bitemporal hemianopsia. The presumed diagnosis was normal pressure hydrocephalus, and his neurologist decided to do a lumbar puncture that revealed an elevated cerebrospinal fluid (CSF) pressure of 30 cm H_2O. An MRI was subsequently obtained (**Fig. 29.8**).

■ Anatomical and Therapeutic Considerations

This patient had two symptomatic lesions, a colloid cyst blocking the foramen of Monro (more on the left side) with obstructive biventricular hydrocephalus, and a pituitary adenoma with compression of the optic chiasm (**Fig. 29.8**). His laboratory data revealed that the adenoma was nonfunctioning but associated with partial hypopituitarism.

Options for Approach

As the two lesions appeared close together, it was tempting to consider not only a single operation, but a single approach, to eliminate both of them. One option would be to remove the adenoma through a transsphenoidal approach, and after that, intentionally open the diaphragma sellae and floor of third ventricle, in an attempt to remove the colloid cyst from below.

Although intuitively attractive, one must immediately recognize that this strategy in fact requires two approaches because in order to prevent the hydrocephalus from causing unpredictable damage by pushing intracranial contents through the opening in the diaphragma, one must start with placement of an external ventricular drain, and that, by definition, adds up to two procedures. Furthermore, unlike a craniopharyngioma which often grows into the third ventricle, the colloid cyst is behind an *intact* tuber cinereum, and opening an anatomically normal floor of the third ventricle wide enough to access the colloid cyst may cause hypothalamic injury. Finally, even if this access was achieved, the cyst attachment to the tela choroidea would be hidden from the surgeon's line of sight from the transsphenoidal route.

Approach of Choice

For all these reasons, we considered the "single approach" transsphenoidal option inadvisable. Instead, since the cyst was

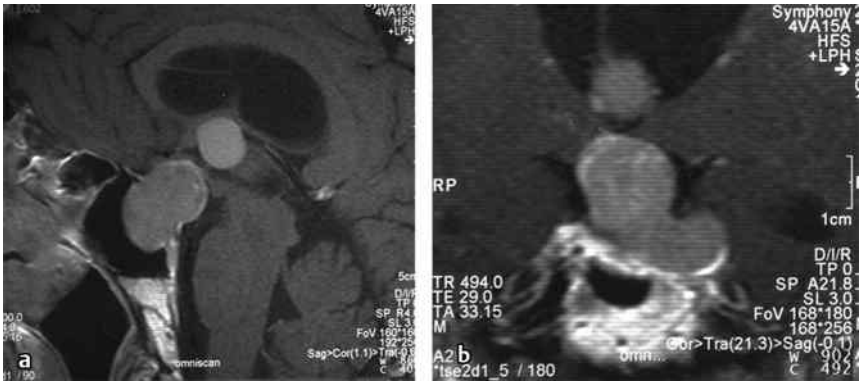


Fig. 29.8 Preoperative MRI. (a) Sagittal and **(b)** coronal images showed that the patient had two simultaneous lesions, a colloid cyst which blocked the foramen of Monro, causing hydrocephalus, as well as a pituitary macroadenoma, with compression of the optic chiasm. The signs and symptoms indicated that both lesions were symptomatic.

eccentric to the left, we chose a left frontal burr hole approach for endoscopic removal of the colloid cyst. This was followed by an endoscopic transnasal removal of the pituitary adenoma in the same session.

Description of the Technique

The patient was positioned supine, the neck flexed 20 degrees, with the head in a neutral position and fixed in a vacuum pillow. A left-sided 4-cm straight incision was made frontally, followed by a burr hole 8 cm from the nasion and 4 cm paramedian (**Fig. 29.9**). For this patient, this was still within the hairline.

An endoscope (rigid lenscope, 6 mm diameter, 2.2 mm working channel, 0 and 30 degrees optic) was introduced through a split-trocar into the left lateral ventricle, immediately revealing the large cyst blocking the foramen of Monro. The cyst wall was covered by the choroid plexus, and on probing, appeared rather firm. The choroid plexus was coagulated with bipolar diathermy and then cut with a sharp scissor. With closed cup forceps the remaining part of the choroid plexus was "scraped off" the cyst wall. Small vessels on the cyst wall were coagulated. The endoscopic perforator was used to open the cyst wall with slight turning movements without applying suction.

The content was viscous with some solid parts. The cyst wall was grasped with forceps and first moved laterally, then backward, followed by an upward movement by rotating the grasping forceps both clockwise and counterclockwise. That maneuver was repeated several times, and with this, the cyst slowly but steadily detached from the tela choroidea. A balloon catheter was placed into the third ventricle below and behind the cyst wall. Inflating the balloon prevented any blood from entering the third ventricle and kept the cyst better positioned inside the foramen of Monro. This move helped to expose the attachment of the cyst at the tela, which was coagulated and then cut. The cyst was too big to remove through the working channel, and so it was held close to the lens while the endoscope was retracted (**Fig. 29.10**).

The endoscope was reinserted, and a small point of bleeding from the tela was detected. Since it did not stop with a minute of continuous irrigation, the balloon catheter was again inflated and pressed against the bleeding part for hemostasis. The endoscope was moved through the foramen into the third ventricle to check the aqueduct and to ensure that no debris or blood was obstructing the aqueduct (**Fig. 29.11**). A triangular shaped piece of Gelfoam was used for the closure, partly pushed into the cortical channel while its larger part is covering the burr hole. Then, the pericranium and galea were closed carefully.

The patient's head was then repositioned into a fully neutral position and then redraped. An endoscopic binostril transnasal-transsphenoidal approach was used to open the sphenoid

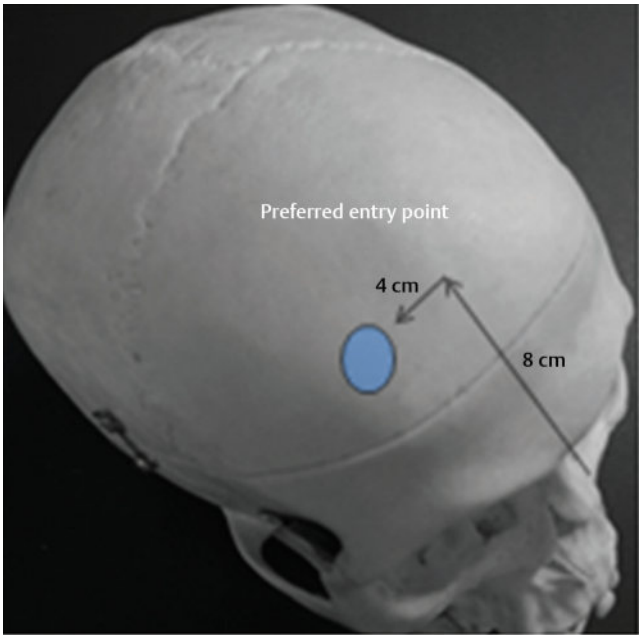


Fig. 29.9 Burr hole placement. The burr hole for the endoscopic approach to the colloid cyst was placed 8 cm from the nasion and 4 cm left of midline.

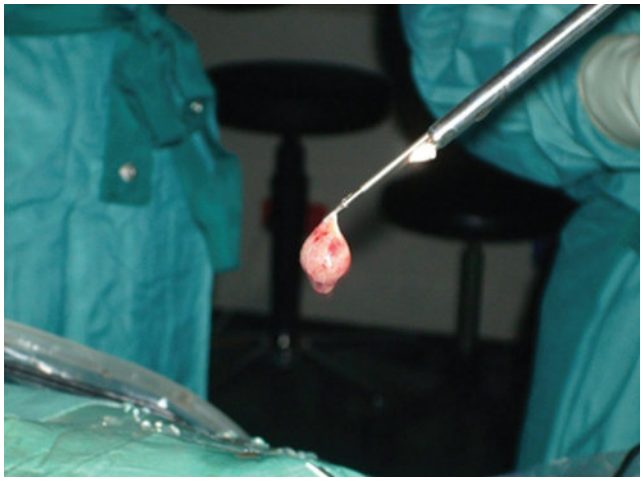


Fig. 29.10 Cyst removal. The colloid cyst was detached from the tela choroidea. Since it did not fit through the working channel, it was removed by retracting the endoscope.

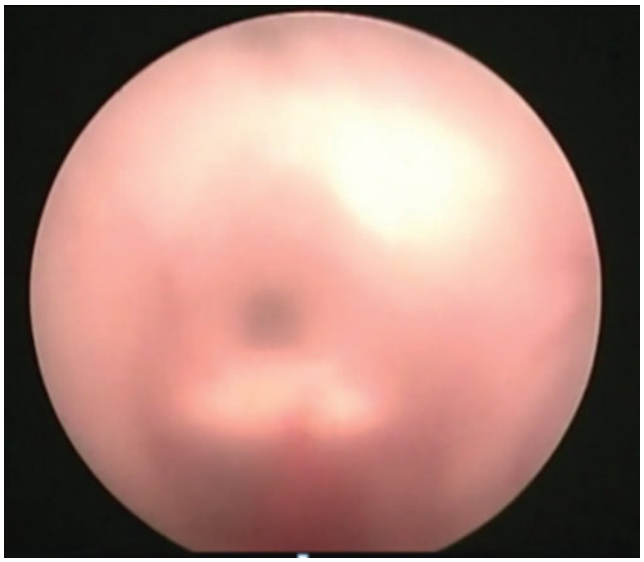


Fig. 29.11 Third ventricle. After removal of the colloid cyst, the entire third ventricle was examined thoroughly with the endoscope to ensure that no debris or blood was obstructing cerebrospinal fluid flow, especially at the aqueduct.

sinus widely, exposing the enlarged sella. After opening the sella widely, exposing the intercavernous sinus superiorly and inferiorly, and the edges of the cavernous sinus bilaterally (i.e., "four blue lines"), the dura was incised and partially removed. A soft adenoma was exposed and removed with suction and curettes. An attempt for extracapsular removal was made, but no clear pseudocapsule could be found. On the left side, the cavernous sinus was completely compressed (but not invaded) by the adenoma. After removal of that part, some holes were identified in the medial wall of the cavernous sinus. Some venous oozing was stopped by application of FloSeal. The sellar diaphragm came down into the sella. The remaining space inside the enlarged sella was filled with some fibrin glue. The floor was reconstructed with parts of the posterior septum that were removed during the initial exposure.

Aftercare

The patient's postoperative course was uneventful. His vision improved, as did his gait, memory, and urinary symptoms. He remained on thyroid supplementation and received testosterone replacement (he stopped this by himself after 7 years). His adrenal axis remained intact. He is now 84 years old and still doing well with no recurrence of either the pituitary adenoma or the colloid cyst (**Fig. 29.12**).

Commentary

In the first section of this chapter, the authors chose to follow a logical path to eliminate the prenidal aneurysm first, and then remove the colloid cyst. There is a rapid decrease in intracranial pressure when a large amount of CSF is released after opening the ventricles. This kind of drop in ICP would put the AVM, and especially the AVM-associated aneurysm, at risk for bleeding. The authors could have also chosen to fully eliminate the aneurysm and AVM first, and then approach the colloid cyst at a later stage.

Open surgical approaches, especially via the transcallosal route, have the disadvantage that the trajectory is in a vertical line from the skull to the foramen of Monro, which, though it provides good exposure to the anterior part of the third ventricle, makes it difficult to access the origin of the colloid cyst, more posteriorly at the roof of the third ventricle. An interforniceal route into the third ventricle could improve this exposure, but places the patient at risk for memory deficits.

Endoscopic removal of the cysts through a frontal burr hole is a less-invasive alternative which is gaining wide acceptance. Its main advantage, aside from smaller incision, less postoperative pain, and shorter hospital stay, is that it brings illumination directly to the region of interest. The technique is straightforward when hydrocephalus expands the foramen of Monro, but when the cyst and foramen are small, in normal-to-small ventricles (**Fig. 29.13a**), the endoscopic transchoroidal approach remains an option.

The Technique of Endoscopic Transchoroidal Approach

In this approach, the anterior septal vein is first coagulated and detached from the choroid plexus. The foramen of Monro can then be enlarged by opening between choroid plexus and thalamostriate vein. Then, by coagulating and cutting the taenia fornices, the choroidal fissure is opened medially, allowing access to the roof of the third ventricle (**Fig. 29.13b**).

Both internal cerebral veins come in view, often pushed apart by the cyst. This gives access to the attachment of the cyst at the tela choroidea. By coagulating and cutting this attachment, and with small rotating motions of the grasping forceps, the cyst wall is freed from the tela. This allows the cyst to be removed completely (**Fig. 29.13c**).

There is a concerning trend in the current literature, when describing cases with small or normal-sized ventricles, of utilizing "gentle inflation of the ventricle." This is a risky technique, as it often infuses more fluid than can escape from the ventricles, and can lead to excessive high intraventricular pressure, bradycardia or even cardiac arrest. Currently, we perform this technique only under continuous monitoring of the intraventricular pressure, since we had a case of postoperative Terson's syndrome.

Despite its strengths, the endoscopic approach is technically challenging, and because of that, it is associated with the risk of leaving unintended cyst residual and higher recurrence rates. It is critical to understand the limitations of the technique of working through an endoscope, especially when there is little space in front of the lens. Each surgeon must further analyze one's own limitations in skill and experience to achieve the best possible result for the patient. My own experience was developed from a personal series of 74 patients, treated endoscopically in the past 27 years. The last 58 have ended with total cyst

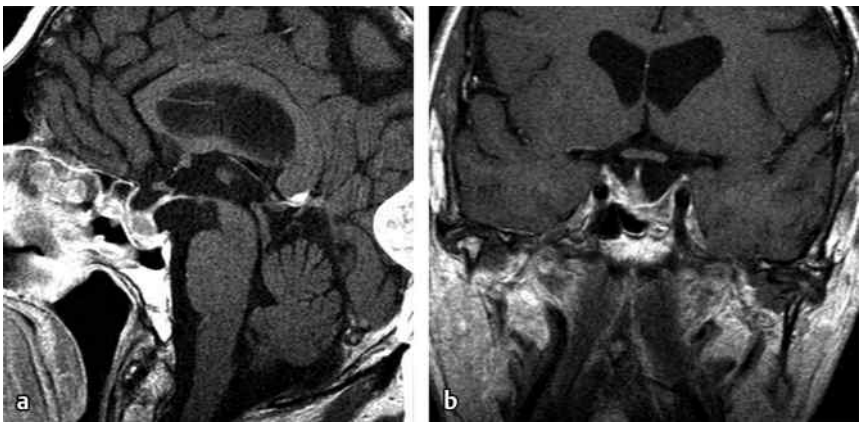


Fig. 29.12 Postoperative MRI. (a) Sagittal and **(b)** coronal images showed no residual of the colloid cyst or pituitary adenoma. There has been no recurrence in more than a decade of follow up.

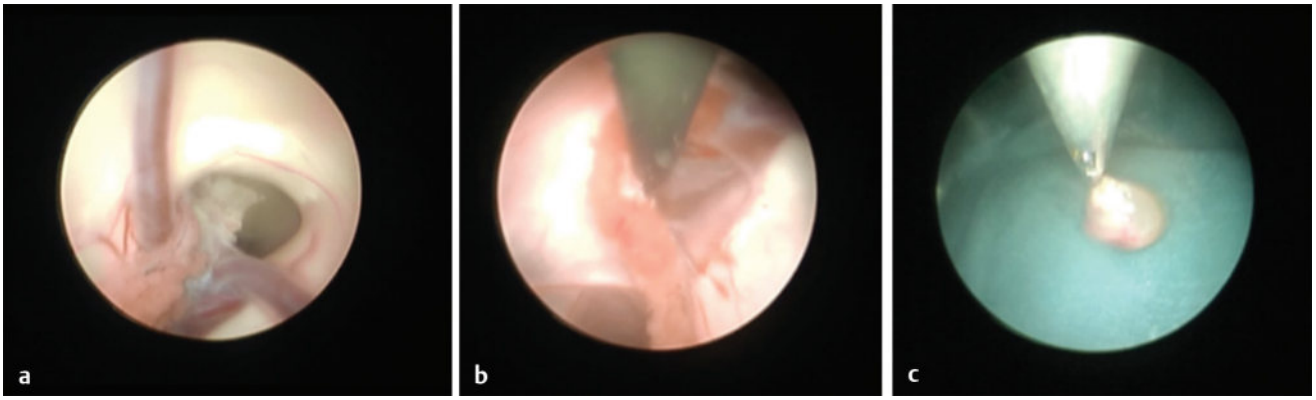


Fig. 29.13 The endoscopic transchoroidal approach. **(a)** Small colloid cyst in normal-sized ventricle without expansion of the foramen of Monro. **(b)** Access into the third ventricle is gained by expanding the foramen of Monro between the choroid plexus and thalamostriate vein, and cutting the taenia fornices. **(c)** Complete removal of the colloid cyst is achieved with rotating movements with the grasping forceps.

removal but this also reflects the rather long and tough learning curve, as most complications and partial removals were in the first 5 years of using the endoscope.

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30 Lateral Ventricle (Atrium)

Tao Xie and Xiaobiao Zhang

Keywords: atrium, trigone, precuneus, optic radiation, interhemispheric fissure, corpus callosum

■ Case Presentation

A 51-year-old woman presented with the chief complaint of headaches for 1 month. The headache was focused on the left occipital side, and she described these headaches as dull. On physical examination, she had trouble seeing on the right side. This deficit was confirmed on visual field testing (**Fig. 30.1**). A magnetic resonance imaging (MRI) with gadolinium was obtained, and it showed a large mass in the atrium of the left lateral ventricle (**Fig. 30.2**).

Questions

1. What MR characteristics of the tumor and the surrounding structures may provide hints to the nature of the mass lesion?
2. What kind of examination and procedure should be done before any operative consideration?

■ Diagnosis and Assessment

The most common lesions that occupy the atrium of the lateral ventricle include meningiomas, choroid plexus papillomas (CPPs), or arteriovenous malformations. Other mass lesions, such as gliomas and metastatic tumors, can also originate in the periatrial area and extend into the atrium. The preoperative MRI of this patient showed a 4 × 3.5 cm homogeneously enhancing mass with smooth margins and moderate surrounding edema (**Fig. 30.3**). A choroid plexus "tail" can be seen on the axial three-dimensional-MPRAGE MR indicating that the arteries feeding the tumor are most likely the choroidal vessels. The rest of the ventricular system appeared normal and there was no sign of hydrocephalus. The differential diagnosis for the tumor includes meningioma, CPP, and choroid plexus carcinoma (CPC). However, a CPP usually has lobulated margins and is frequently associated with hydrocephalus due to overproduction of cerebrospinal fluid (CSF). On the other hand, a CPC usually shows a heterogeneous enhancement pattern, lobulated margins, and significant peritumoral edema with signs of parenchymal invasion. As such, given the MR characteristics of the patient's lesion, the most likely diagnosis was a meningioma.

Peritumoral edema of the meningioma is common, and 38 to 67% of patients with intracranial meningioma present with varying amounts of brain edema. It is well known that glucocorticoids reduce this edema, and we advocate the use of methylprednisolone for at least 48 hours before any surgical intervention.

Preoperative angiography can be of significant diagnostic value. Although a massive tumor 'blush" does not entirely rule out a meningioma, one would expect the choroid plexus tumors (CPP and CPC) to be far more vascular than the usual meningioma. On the contrary, a marginally or moderately vascular tumor on angiography would be highly suggestive of a meningioma. Trigonal meningiomas are usually supplied by the anterior and posterior lateral choroidal arteries. Although infrequently accessible by microcatheters, preoperative embolization, in the rare cases when it is feasible, can greatly facilitate the resection process.

■ Anatomical and Therapeutic Considerations

The atrium of the lateral ventricle is a triangular area that is defined by the temporal horn inferiorly, the occipital horn

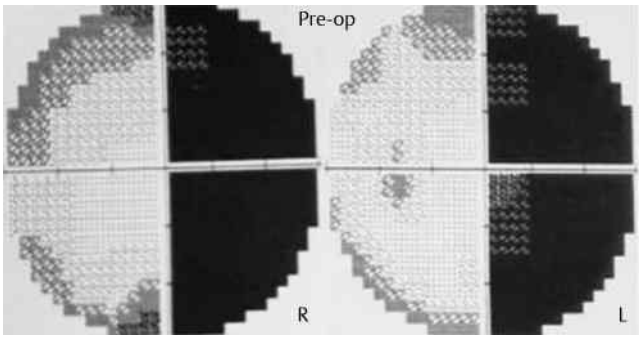


Fig. 30.1 Preoperative visual fields. This examination showed right homonymous hemianopsia.

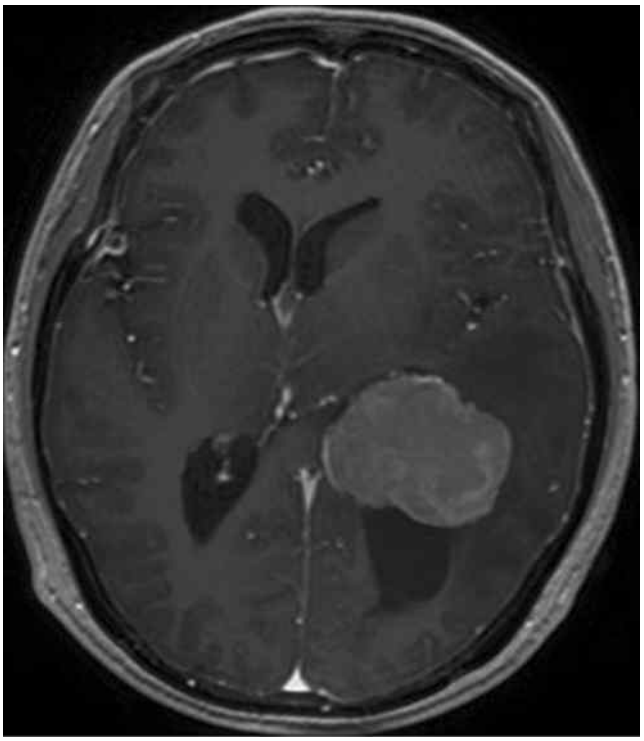


Fig. 30.2 Preoperative MRI. Axial MRI with gadolinium showing a huge mass (4 × 3.4 cm) in the left atrium of the lateral ventricle.

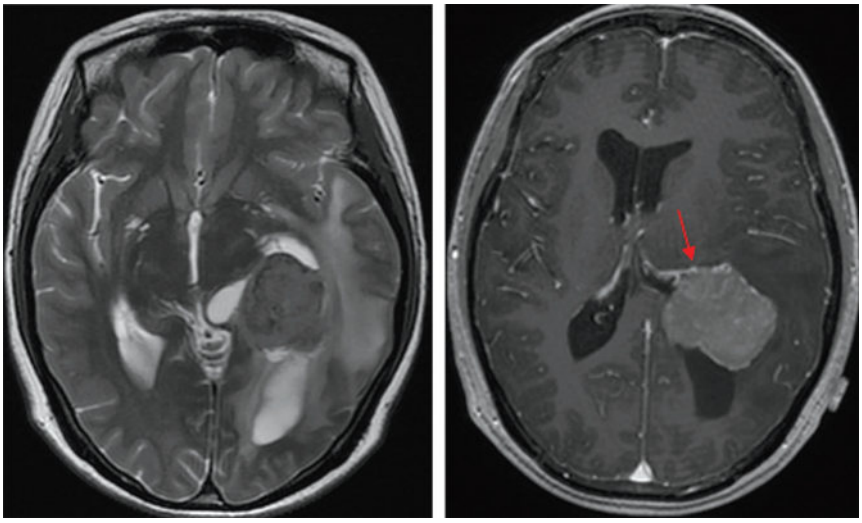


Fig. 30.3 Preoperative MRI. (a) Axial T2 MRI showed the surrounding brain edema and **(b)** the choroid plexus tail sign was clearly seen on the axial three-dimensional- MPRAGE MR (*red arrow*).

posteriorly, and the body of the lateral ventricle anteriorly (**Fig. 30.4**). The inferior wall of the atrium is formed by the collateral eminence, which overlies the collateral sulcus. The medial wall of the atrium and occipital horn is the calcar avis, which overlies the calcarine sulcus. The lateral wall consists of the tapetum. The atrium is intimately related to fibers of the optic radiations, as all three major fiber bundles of the optic radiation (anterior, central, and posterior) run along the edges of the atrium (**Fig. 30.5**). The anterior bundle, commonly known as Meyer's loop, courses posteriorly along the inferior aspect of the trigone and occipital horn. The central bundle extends posteriorly over the lateral wall of the trigone and occipital horn, before terminating at the occipital pole. The posterior bundle passes over the superolateral aspect of the trigone and terminates posteriorly on the superior bank of the calcarine sulcus. Taken together, the entire lateral and inferior wall of the trigone is covered by fibers of the optic radiations. In contrast, the medial wall of the trigone is entirely free of optic radiation fibers.

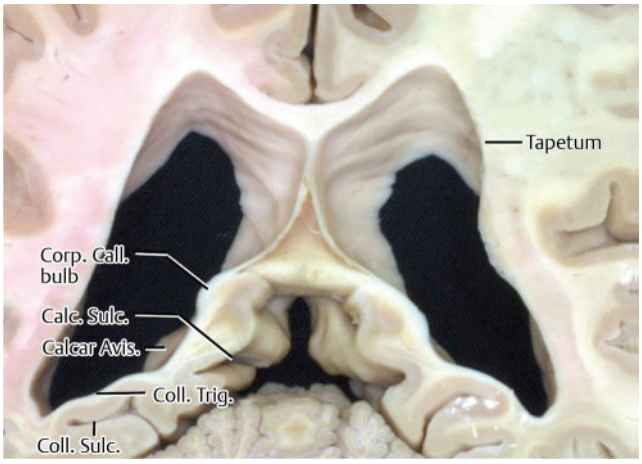


Fig. 30.4 Structures forming the surfaces of the atrium of the lateral ventricles. Calc., calcarine; Call., callosum; Coll., collateral; Corp., corpus; Sulc., sulcus; Trig., trigone. (Reproduced from Torres-Corzo J, Rangel-Castilla L, Nakaji P, eds. Neuroendoscopic Surgery. 1st ed. Thieme; 2016.)

Options for Approach

There are many potential approaches to the atrium of the lateral ventricle, but the ideal approach should provide the shortest working distance and largest working angle to the center of the lesion without jeopardizing the functional cortex or white matter tracts. The superior parietal lobule approach, the transtemporal approach, or the lateral temporoparietal approach all provide direct access to the atrium (**Fig. 30.6**). Of these, the superior parietal lobule approach is probably used most frequently for tumors of the atrium. The main disadvantage of this approach is that the surgeon has to traverse a large amount of normal brain in the superior parietal lobe to reach the target. The resulting working distance to the target is long, and in addition, the damage to the parietal lobe tissue can cause a wide spectrum of neurological deficits from visual impairment to Gerstmann's syndrome. The situation is worse if the parietal lobe was already affected by peritumoral edema before surgery. Furthermore, when the superior parietal lobule approach is used for meningiomas of the atrium, the tumor mass blocks the surgical access to the arterial supply. Blood loss may be significant as the tumor must be debulked sufficiently before the feeding vessels behind it can be eliminated. Similarly, the transtemporal and lateral temporoparietal approaches are associated with significant risks of postoperative neurological morbidity, not only because of potential damage to the visual fibers, but also because of the chance of postoperative seizures.

The posterior approaches (including the posterior transcallosal and the interhemispheric parieto-occipital approach) may avoid the optic radiation surrounding the roof and the lateral wall of the atrium, but the working corridors with these approaches are quite narrow. In the specific case of our patient, the ipsilateral posterior interhemispheric approach is worth consideration. However, there would be a right-angled turn from the surgeon's line of sight to reach the target, and as such, one would have to forcefully retract the already edematous ipsilateral hemisphere to gain sufficient working space.

Approach of Choice

In order to avoid injury to the swollen ipsilateral parietal lobe, we chose to utilize another surgical corridor, via the

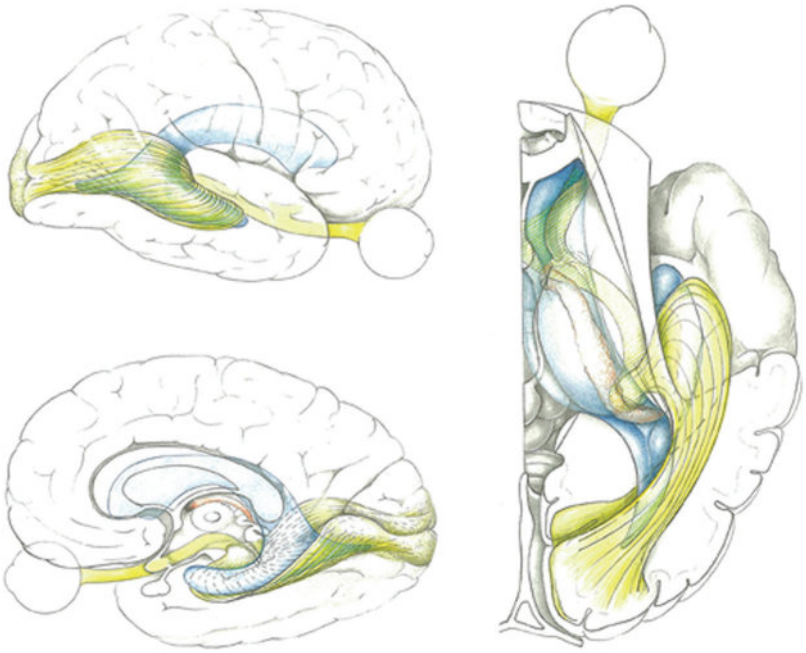


Fig. 30.5 The pathway of optic radiation. Lateral, medial, and superior view of the fibers of the optic radiation, showing that the superior and lateral walls of the ventricular trigone are blanketed by optic fibers. (Reproduced from Yaşargil M, Curcic M, Teddy P et al., eds. Microneurosurgery. Volume III B. AVM of the Brain, Clinical Considerations, General and Special Operative Techniques, Surgical Results, Nonoperated Cases, Cavernous and Venous Angiomas, Neuroanesthesia. 1st ed. Thieme; 1998.)

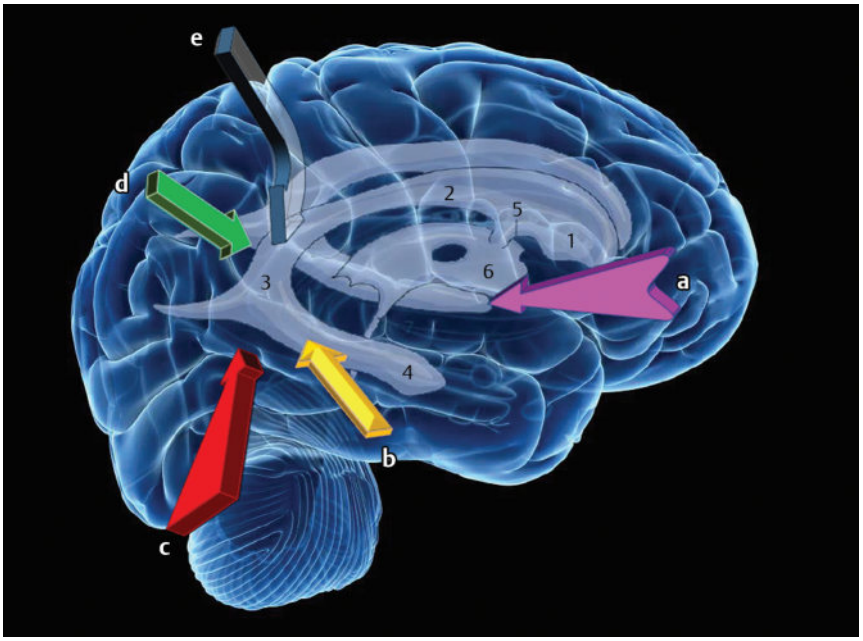


Fig. 30.6 Approach options to the atrium of the lateral ventricle. (a) Distal sylvian approach; **(b)** transtemporal approach; **(c)** supracerebellar transtentorial approach; **(d)** superior parietal lobule approach; **(e)** posterior interhemispheric transcallosal or transprecuneus approaches, which can be performed either from the side ipsilateral or contralateral to the tumor. 1, frontal horn; 2, cella media; 3, atrium of lateral ventricle; 4, temporal horn; 5, foramen of Monro; 6, third ventricle.

contralateral transfalcine transprecuneus route. In 1995, Goel first described the transfalcine approach to contralateral hemispheric tumors and found that it provided a wide exposure without the need for traversing or retracting vital cerebral cortex. Subsequently, both laboratory and clinical studies have confirmed the feasibility and safety of this approach. Instead of traversing the superior parietal or temporal lobe to reach the atrium of the lateral ventricle, this approach goes through the precuneus. The precuneus is a part of the medial parietal lobe and is bounded anteriorly by the marginal branch of the cingulate sulcus, posteriorly by the parieto-occipital sulcus, and inferiorly by the subparietal sulcus (**Fig. 30.7**). Its main function is in visuospatial processing, episodic memory, self-awareness, and some aspects of consciousness. Because of the nature of these functions, injury to this region leads to deficits which are either temporary or difficult to detect.

Compared to the ipsilateral interhemispheric approach, both laboratory and clinical studies have indicated that the contralateral approach improves the exposure by providing a more direct trajectory from surgeon to target (**Fig. 30.8**). Furthermore, in cases where there is significant parietal lobe edema, not only does a contralateral approach eliminate the need to retract the swollen brain, but the edema actually assists the resection as it "pushes" the tumor medially into the surgeon's field. A medial-to-lateral approach through the precuneus also enables the surgeon to eliminate the choroidal blood supply to the tumor early in the operation, thus significantly reducing the blood loss during surgery.

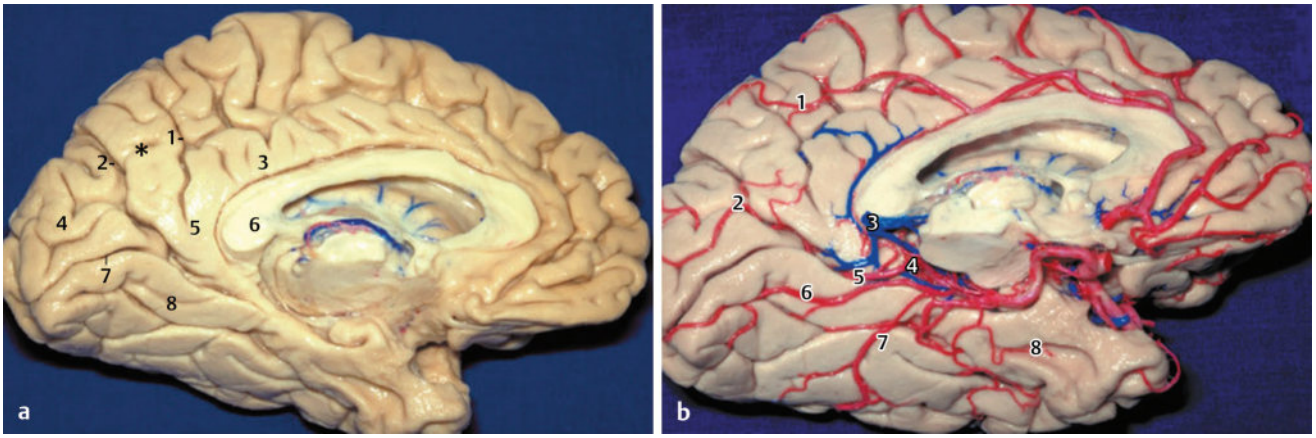


Fig. 30.7 Midline views of the left hemisphere. (**a**) Cortical structures: *, precuenus; 1, subparietal sulcus; 2, parieto-occipital sulcus; 3, cingulate gyrus; 4, cuneus; 5, isthmus of the cingulate gyrus; 6, splenium of the corpus callosum; 7, calcarine sulcus; 8, lingual gyrus. (**b**) Vascular structures: 1, branches from the ACerA; 2, branches from the parieto-occipital artery; 3, vein of Galen and its tributaries; 4, P3 segment of the posterior cerebral artery; 5, P4 segment of the posterior cerebral artery; 6, inferior temporal artery (posterior group); 7, inferior temporal artery (intermediate group); 8, inferior temporal artery (anterior group). The precuneus is relatively devoid of vascular structures.

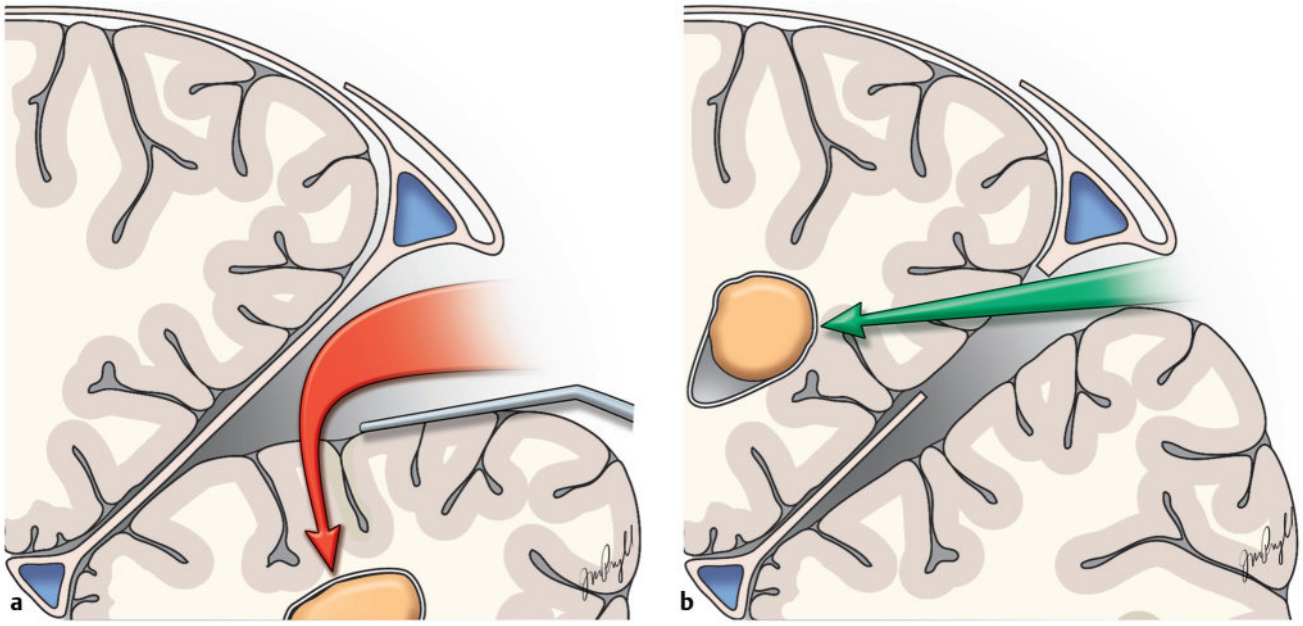


Fig. 30.8 Comparison between approaches through the precuneus. **(a)** Ipsilateral and **(b)** contralateral approaches are shown. The contralateral approach provides the surgeon with a more direct trajectory and better exposure without excessive retraction.

However, every coin has two sides. The main risk of the contralateral transfalcine transprecuneus approach is bihemispheric injury as it places the healthy hemisphere at risk. This is particularly relevant when bridging veins from the parietal lobe to the superior sagittal sinus (SSS) are encountered before the dissection of the interhemispheric fissure. If the ipsilateral parietal lobe is affected by edema, venous infarction of the contralateral, healthy parietal lobe may have catastrophic consequences. Fortunately, bridging veins posterior to the vein of Trolard are sparse, making venous infarction rare for this approach. With upper body elevation to facilitate venous drainage, mannitol and hyperventilation to reduce intracranial pressure, and natural gravity retraction to provide adequate operating space, risk to the contralateral hemisphere can be minimized.

With all those considerations made, the plan was to fully resect the patient's tumor via the contralateral transfalcine, transprecuneus approach.

Questions

1. What kind of the patient position will you choose? How will the position impact on this approach?
2. What is the function of the precuneus? Is this area truly silent?
3. What kind of extension of the atrium lesions will be most suitable for the contralateral transfalcine transprecuneus approach?

Description of the Technique

After induction of general endotracheal tube anesthesia, the patient was placed in the prone position. The upper body and head were elevated 30 degrees, and her head was placed in a three-pin fixation and turned approximately 15 degrees to the side ipsilateral to the lesion. This allowed gravity to retract the contralateral parieto-occipital lobe. Registration of the image guidance system was performed (see **The Three Approach Elements**).

The Three Approach Elements

Corridor: interhemispheric

Craniotomy: parieto-occipital

Modifiers: incision of the falx

An 8-cm straight-line skin incision was made about 1 cm inferior to the lambdoid suture and perpendicular to the sagittal suture. A contralateral parasagittal parieto-occipital craniotomy, approximately 4 × 4 cm, extending across the sagittal sinus, was performed. The dura mater was opened in a semicircular fashion with the base reflected toward the SSS, and CSF was released from the interhemispheric fissure for brain relaxation. Arachnoid adhesions near the SSS were carefully dissected until the falx was identified. Dissection into the interhemispheric fissure then proceeded with caution to preserve any bridging veins (see **Operative Setup**).

The splenium of the corpus callosum was identified, and cutting the falx was the essential next step in the approach. The anatomic study of the venous plexus within the falx cerebri suggests that most falcine veins reside in its posterior one-third of the falx cerebri. In addition, most of these drain into the straight sinus and are therefore concentrated in the inferior portion. An arc-shaped incision can avoid this falcine venous plexus (**Fig. 30.9**). MR venography neuronavigation, endoscopic examination, and fluorescein dye can all help to avoid these veins.

For our patient, the falx was cut parallel to the line that was 1 cm inferior to the SSS, and the dura was opened in an arc-shaped fashion, extending from the SSS toward the straight sinus posteroinferiorly and the inferior sagittal sinus anteroinferiorly. Three-dimensional MR venography neuronavigation and mini-Doppler imaging were used to confirm the location of the sinuses.

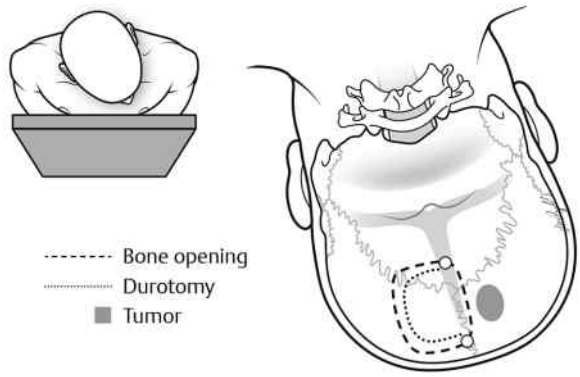
Operative Setup

Position: prone, head turned towards the ipsilateral side

Incision: straight-line incision perpendicular to the sagittal suture

Bone Opening: contralateral parasagittal parieto-occipital craniotomy

Durotomy: "U-shaped" based on sagittal sinus



The precuneus gyrus on the target side was then identified with neuronavigation, using the parieto-occipital sulcus as a landmark. A longitudinal corticotomy was made in the precuneus gyrus to expose the medial side of the tumor (**Fig. 30.10**). After debulking of the tumor, the blood supply from the choroid plexus was exposed and eliminated. Finally, the capsule was dissected, and the tumor was completely removed. The neuroendoscope provided a wider viewing-angle for revealing the inferior sagittal sinus and straight sinus, removing the intraventricular clot, as well as inspecting for residual tumor

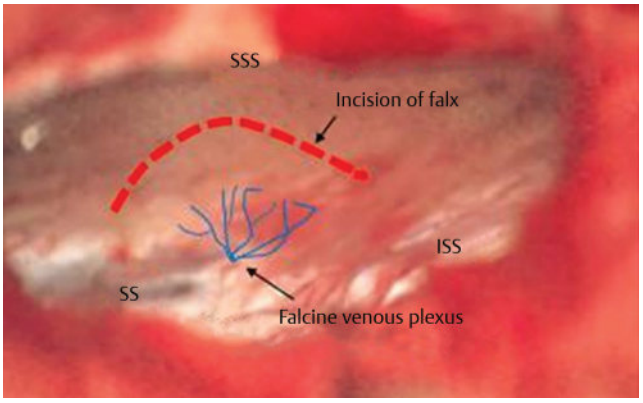


Fig. 30.9 Diagram depicting the view of the falx during the contralateral transfalcine transprecuneus approach. The planned arc-shaped incision of the falx (*dotted line*) is shown. ISS, inferior sagittal sinus; SS, straight sinus; SSS, superior sagittal sinus.

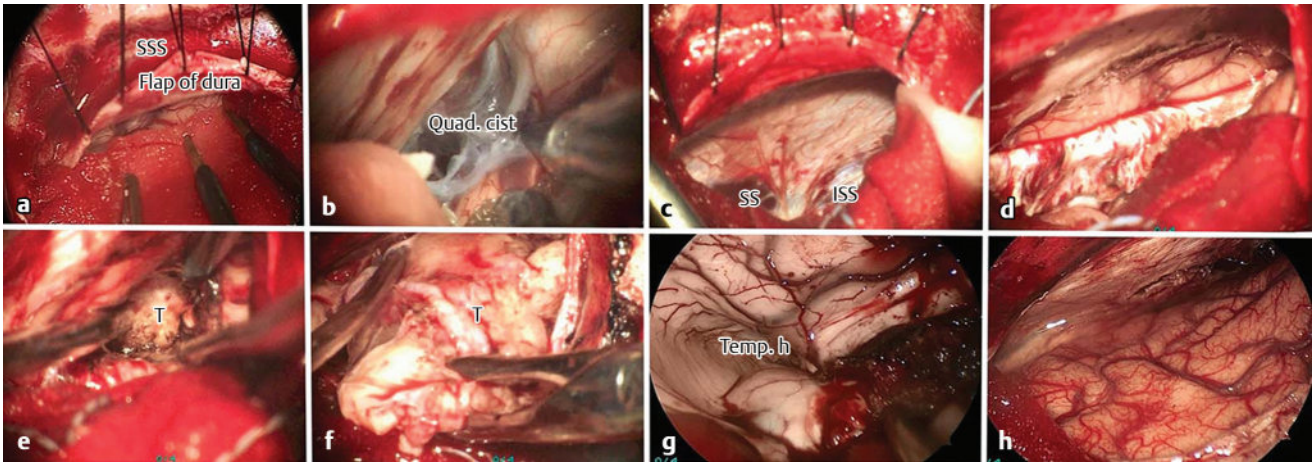


Fig. 30.10 Intraoperative images. (a) Endoscopic view of the bone area and dural flap. **(b)** The quadrigeminal cistern was opened to release the cerebrospinal fluid. **(c)** Microscope view of the inferior sagittal sinus and straight sinus. **(d)** After cutting the falx, the contralateral precuneus gyrus was located. **(e)** The medial side of the tumor was seen after the longitudinal corticotomy in the precuneus gyrus. **(f)** After dissecting the border of the tumor, the capsule of the tumor was completely removed. **(g)** The endoscope provided supplemental visualization of the temporal horn to check for any residual tumor and to remove the iatrogenic clot. **(h)**. The flap of the falx was sutured after the hemostasis. ISS, inferior sagittal sinus; Quad. Cist, quadrigeminal cistern; SS, straight sinus; SSS, superior sagittal sinus; T, tumor; Temp. h, temporal horn.

and bleeding (**Fig. 30.11**). The flap of the falx was closed with continuous sutures, and then the dura mater, bone flap, galea, and skin were closed layer by layer.

Surgical Pearls

1. Patient position and gravity retraction: The prone position with a 30-degree body elevation offers optimal venous drainage and reduces the intracranial pressure. The 15-degree rotation facilitates retraction of the parieto-occipital lobe by gravity. The force of gravity also helped in delivering the tumor into the surgeon's operative field. Some surgeons have advocated using the lateral position, with the head rotated towards the floor, which would achieve the same gravity retraction. The choice between the two is up to the surgeon's habit.
2. Craniotomy and bridging veins: A contralateral parasagittal parieto-occipital craniotomy extending across the sagittal sinus minimizes the degree of retraction. The bridging veins in the parieto-occipital region tend to join the SSS before the lambdoid suture. Therefore, it is safer to perform craniotomy behind the lambdoid suture.
3. Dural incision: A "shallow" dural opening over the convexity is preferred because it minimizes the chances of the brain being injured, or strangulated, along the free edge of the dural incision. Some surgeons advocate basing the "U" shape *laterally*, and cutting the dura near the SSS to avoid this problem.
4. Falx incision: Injuring the straight sinus during this step can lead to massive bleeding. It is therefore critical to identify this structure before proceeding with this step. Fluorescein/indocyanine green may be helpful to visualize the straight sinus in some cases.
5. Tumor exposure: The contralateral precuneus gyrus is identified by neuronavigation using the parieto-occipital sulcus as a landmark. If this fails, the precuneus can be identified as the region posterior to the splenium of the corpus callosum and anterior to the parieto-occipital sulcus. The former can be identified by its white color, and the latter is the biggest, widest sulcus in the region.
6. Tumor resection: Most trigonal meningiomas of the lateral ventricle receive blood from the anterior and posterior lateral choroidal arteries. After coagulating and incising the blood supply, the bleeding is reduced during the resection. Preoperative angiography may supply more accurate information on the blood supply. When the internal debulking is done, brain cotton is placed anterior to the tumor to prevent the blood flow entering the ventricular system from the surgical field. Minimizing the coagulation of the choroid plexus may prevent injury to visual function by preserving the subependymal arteries that feed the optic radiation.
7. Neuroendoscopic-assisted procedure: The endoscope provides supplemental visualization of any residual tumor and should be routinely used to look around corners after the main resection is complete.

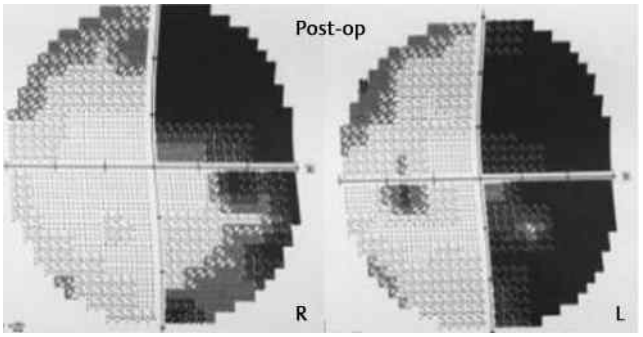


Fig. 30.11 Postoperative visual fields. This examination showed improvement in the right homonymous hemianopsia after the operation.

■ Aftercare

The patient tolerated the surgery well with no new neurological issues. Her steroid therapy was continued for 4 days, and

she was discharged 7 days after the surgery. The postoperative pathological diagnosis was fibrous meningioma (WHO I), and the postoperative visual field examination showed improvement in the homonymous hemianopsia (**Fig. 30.11**).

■ Possible Complications and Associated Management

Aside from the intraoperative risk of bleeding from venous sinuses, the main complications after this procedure are venous infarction, intraventricular hemorrhage, and hydrocephalus. To avoid injury to the healthy contralateral hemisphere, preservation of bridging veins has already been mentioned. It is important for the craniotomy opening to be wide enough in the anterior–posterior dimension, such that if bridging veins are encountered, the dissection into the interhemispheric fissure can proceed in a gap between these veins.

Significant postoperative ventricular hemorrhage would require immediate reoperation. The only ways to prevent this are meticulous hemostasis around the tumor bed and stringent blood pressure control after the operation. An external ventricular drain, placed in the tumor bed after the operation, can remove small amounts of blood and surgical debris and should be liberally used at least for several days postoperatively. Theoretically, this would reduce the chances of hydrocephalus, as it is less likely for debris to block the arachnoid granulations. Long-term complications include entrapment of the temporal horn, which can be treated with endoscopic lysis of adhesions, or shunting.

Perspective

Ignatius N. Esene, Omer S. Sahin, and Mustafa K. Baskaya

■ Introduction

Intraventricular tumors in general and atrial tumors in particular, are notorious for posing difficulties for neurosurgeons because of their deep and delicate location, as well as their rarity. With the myriad of approaches available, achieving balanced mastery of all the operations is difficult. That said, we should be familiar with all of them, and, as usual, an intimate knowledge of the surrounding critical anatomy is the first step to our learning.

Surgery is the mainstay in the treatment for both *true* intraventricular tumors, such as meningioma, which originate in the atrium, as well as periventricular tumors, such as gliomas, which grow into the atrium. Total resection is generally the goal. The ventricular atrium can be accessed via direct transcortical or the interhemispheric corridor, but neither is risk free. As discussed in the previous section, the atrium is surrounded laterally and inferiorly by the optic radiation, and transcortical approaches, whether they traverse the temporal or parietal cortex, risk injury to visual function. The interhemispheric route, which includes the posterior interhemispheric pericallosal or transcallosal, and the parieto-occipital approaches, evades the optic radiations, but is limited by the long operative distance. The approach discussed in the first section of this chapter, the interhemispheric transfalcine transprecuneus approach, also poses the additional risk of damaging the contralateral occipital lobe from retraction injury. Variations in the anatomy of the subparietal and parieto-occipital sulcus can make the localization of the precuneus difficult, and transgressing the precuneus may not in fact be harmless, since postoperative memory disturbance has been reported in some cases.

For all these reasons, selecting an "ideal" approach for an intraventricular atrial lesion remains challenging.

■ Case Presentation 1

A 68-year-old woman presented with congruent left homonymous hemianopsia. Her MRI showed a right-sided, 5-cm, contrast-enhancing intraventricular mass consistent with a meningioma (**Fig. 30.12**). The tumor involved the right choroid plexus, and together with the vasogenic edema, exerted significant mass effect on the right lateral ventricle.

Anatomical and Surgical Considerations

The lesion occupied almost the entire right ventricular atrium and extended into the occipital horn. Compression of the optic radiation explained the visual field deficits. Although a posterior interhemispheric approach seemed to be an ideal route, this would result in a long operative trajectory. Significant retraction of the calcarine cortex would be needed to reach the lateral and anterior aspect of the tumor, and with the existing edema, the risk to further visual damage was high. The shortest distance to the tumor was through the temporal cortex, and we opted for the transtemporal approach through the middle temporal gyrus (**Fig. 30.12**).

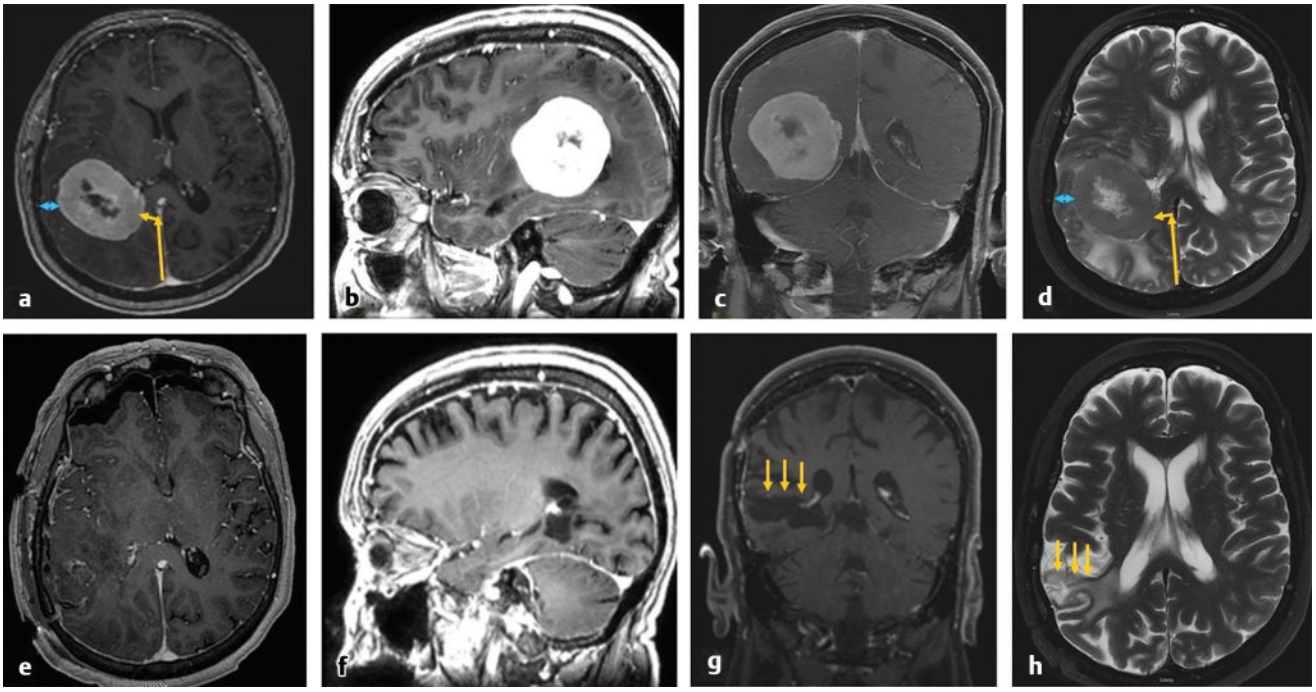


Fig. 30.12 Case 1 images. Preoperative T1-postcontrast **(a)** axial, **(b)** sagittal, and **(c)** coronal MRI showed a very large right intraventricular meningioma of the atrium. **(d)** T2 MRI showed surrounding edema. *Blue arrows* show the short operative distance for the transtemporal approach and the *yellow arrow* longer operative corridor for the interhemispheric approach **(a, d)**. **(e-h)** Postoperative MRI shows the transtemporal approach (*yellow arrows*) and complete tumor resection.

Description of the Technique

The patient was placed in the supine position, with head fixed and turned to the left. A curvilinear incision was made followed by a temporal craniotomy, and the dura then opened in a C-shaped fashion. The neuronavigation was used to identify the transcortical entrance in the middle temporal gyrus. One of the sulci leading to the tumor was carefully dissected, and white matter was transgressed. The tumor came into view at the ventricular surface. The margins were defined, and the mass was debulked, and this sequence was repeated (**Fig. 30.12**). The tumor seemed to have parasitized the pial arterial supply and devascularization enabled further debulking until we came across the tumor's attachment to the ependymal wall. We then carefully swept the choroid plexus away from the tumor surface at its base and coagulated the feeders from the choroid plexus. The remaining shell of the tumor was removed en bloc. An external ventricular drain was placed into the temporal horn before the closure.

Aftercare

The postoperative course was uneventful, and the MRI showed the transtemporal transsulcal corridor to the right atrium, confirming a total resection (**Fig. 30.12e–h**). The histopathologic diagnosis was a WHO grade I meningioma. The patient's visual symptoms completely resolved and there was no recurrence at two years follow-up.

■ Case Presentation 2

A 63-year-old man presented with progressive difficulty in reading and a warm sensation along the right side of his body. His visual examination revealed a right superior quadrantanopsia. An MRI scan showed a left thalamic mass measuring 4.5 × 3 cm (**Fig. 30.13**).

Anatomical and Surgical Considerations

The tumor expanded the left ventricular atrium with extension into the left posterior hippocampus and cerebral peduncle. Enhancing tissue was noted along its inferolateral margin with a punctate area of restricted diffusion suggestive of increased cellularity. Approach options to this tumor include the interhemispheric transcallosal and superior parietal lobule approaches. However, a large sulcus was noted right over the tumor, and this would provide a relatively short trajectory to the tumor. As such, we elected to use a transcortical approach through this large sulcus (**Fig. 30.13d**).

Description of the Technique

A craniotomy over the large sulcus was performed in a supine position with the aid of neuronavigation. Following a C-shaped dural opening, we dissected the large sulcus overlying the tumor and used 10–0 pial–arachnoid retraction sutures to broaden the exposure (**Fig. 30.14**). Once the bottom of the sulcus was reached, a small corticectomy was made to enter the atrium. The part of the tumor that was encountered first was the contrast-enhancing portion, and it had a distinct purplish hue. As we removed this part, we came across a grayish portion which corresponded to the nonenhancing portion. Motor evoked potentials were checked and remained unchanged throughout the case. After a total removal of the enhancing portion and subtotal resection of the nonenhancing part of the tumor, we decided to stop since we were close to the internal capsule.

Aftercare

The patient woke up with mild right hemiparesis which improved over a few days. Postoperative MRI confirmed gross total resection of enhancing component (**Fig. 30.15**). Histopathologic examination revealed a WHO grade IV glioblastoma.

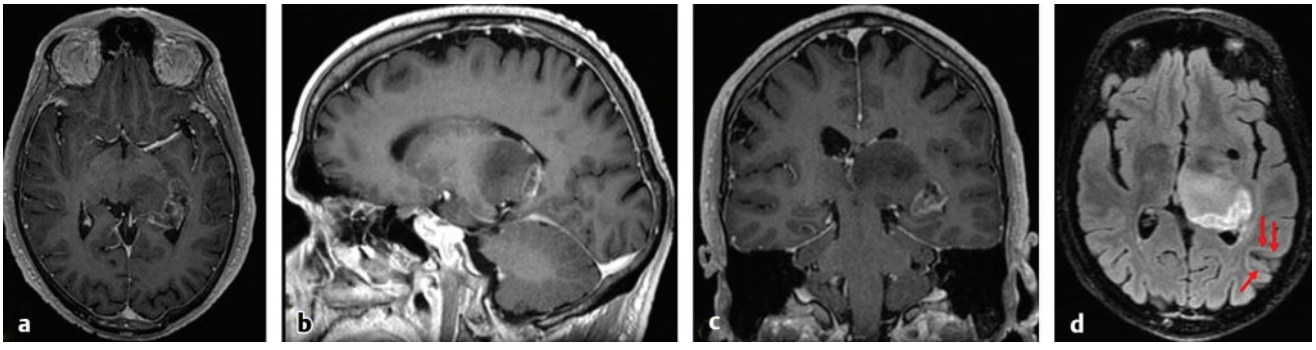


Fig. 30.13 Preoperative images, Case 2. T1-weighted postcontrast **(a)** axial, **(b)** sagittal, and **(c)** coronal images showed a left thalamic tumor with intraventricular extension into the atrium. **(d)** CUBE image showed a large sulcus with atrophic gyri which served as the route of entry (*red arrows*).

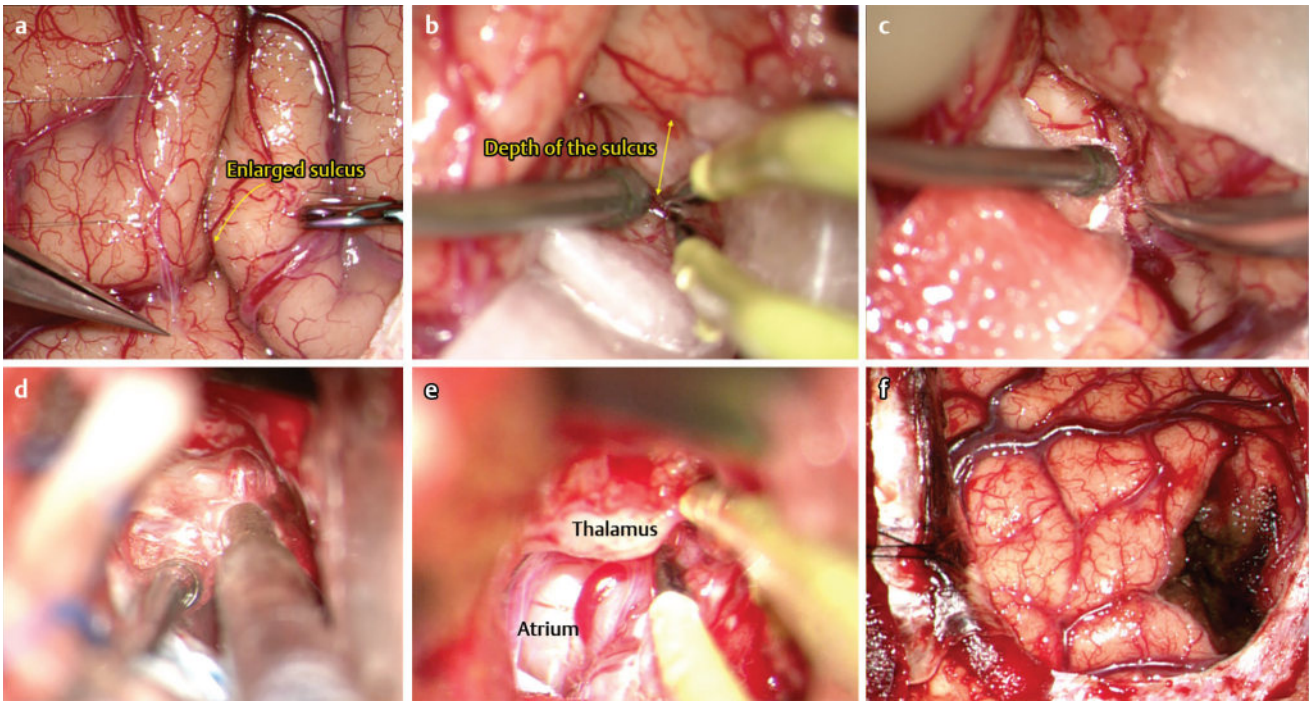


Fig. 30.14 Intraoperative images, Case 2. (a) After opening the dura, microscopic overview of the enlarged sulcus is demonstrated. **(b, c)** After wide transsulcal dissection, the depth of the sulcus has been reached. **(d)** Debulking of the tumor with the aid of an ultrasonic aspirator. **(e)** Following the tumor resection, view of the thalamus and the atrium of the lateral ventricle. **(f)** Overview before the dural closure.

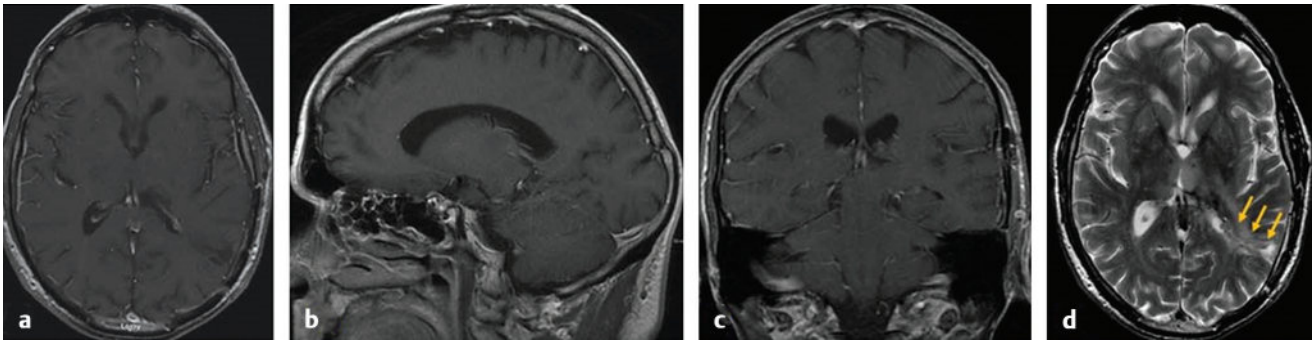


Fig. 30.15 Postoperative MRI images, Case 2. (a–d) Gross total resection of the contrast-enhancing and subtotal resection of nonenhancing portions of the tumor. Note the transtemporal transcortical approach (*yellow arrows in* **d**).

■ Case Presentation 3

A 26-year-old right-handed woman presented with short-term memory disturbances, behavioral changes, and headaches. She had a history of psychiatric illness but the neurological examination was nonfocal. Preoperative MRI scan showed a 6-cm intraventricular mass, consistent with high-grade glioma. There was surrounding vasogenic edema and entrapment of the left lateral ventricle (**Fig. 30.16**).

Anatomical and Surgical Considerations

The tumor involved the splenium of the corpus callosum and extended into the right thalamus and the atrium, causing entrapment of the left lateral ventricle (**Fig. 30.16**). Engorged vessels were noted near the mass, including posterior choroidal and pericallosal branches. Since the tumor involved the ventricle on both sides, the occipital interhemispheric approach was selected since it can provide bilateral access. Cortical veins draining into the SSS and the long surgical corridor were potential obstacles in this approach. Another option would be the superior parietal lobule approach which would offer access to both the medial and lateral walls of the atrium. Albeit "noneloquent," the thick brain tissue that must be crossed to reach the target is the main disadvantage.

Description of the Technique

With the patient in a prone position, a large biparieto-occipital craniotomy was performed. The dura was opened based on the sagittal and transverse sinuses. Following the interhemispheric fissure, we visualized the splenium of corpus callosum, heavily infiltrated with tumor (**Fig. 30.17**). After initial debulking, the tumor was dissected from right intraventricular structures

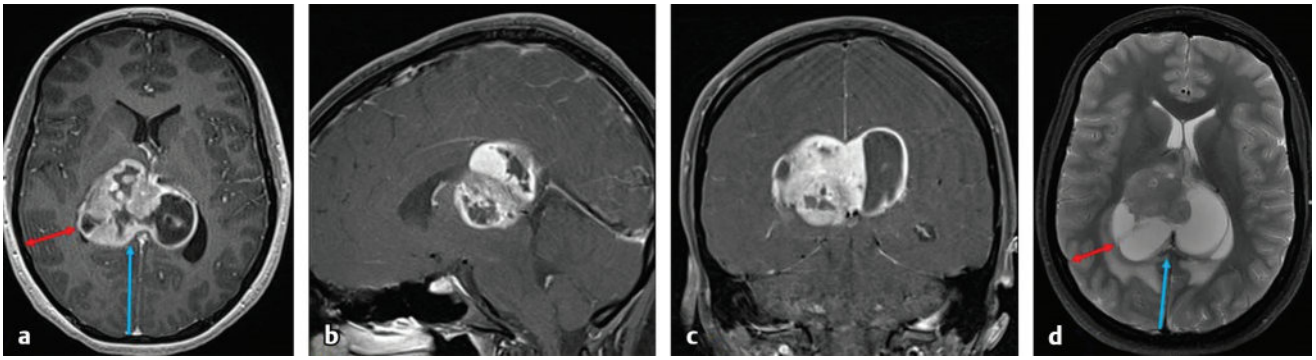


Fig. 30.16 Preoperative images, Case 3. T1-weighted postcontrast images **(a)** axial, **(b)** sagittal, and **(c)** coronal images showed the intraventricular tumor involving the splenium and the thalami. **(d)** T2-fast spin echo image showed peritumoral edema and entrapped left lateral ventricle. *Blue arrow* **(a, d)** shows the interhemispheric corridor and *red arrow* the transcortical approach.

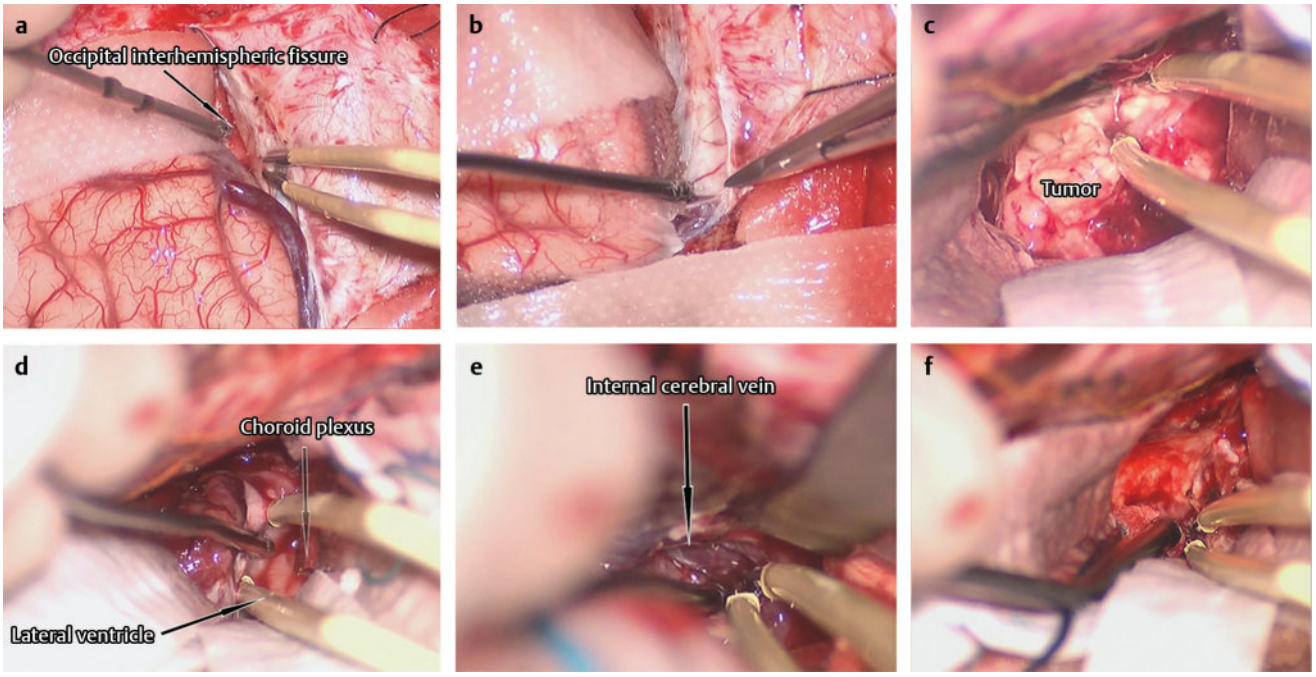


Fig. 30.17 Intraoperative images, Case 3. **(a)** Microscopic overview after reflection of the dura. Note that with the patient positioned prone, her feet are towards the top and vertex towards the bottom of the image. **(b)** Gentle retraction of the midline gyrus to expose interhemispheric fissure. **(c)** Coagulation and debulking of the tumor. **(d)** The close relation of the lateral ventricle and choroid plexus with the tumor. **(e)** Preservation of the internal cerebral vein is demonstrated. **(f)** Overview after the gross total resection of the tumor.

including internal cerebral vein (ICV) and choroid plexus. Next, we debulked the tumor toward the left side. After defining the vein of Galen and both ICV, we gently dissected the tumor off the right ICV to which it was adherent. After releasing tumor attachments at the left choroid plexus and right thalamus, a total resection was accomplished.

Aftercare

She woke up from surgery without new deficits and made an excellent recovery. Postoperative MRI revealed no tumor residual (**Fig. 30.18**). Histopathologic examination was reported as a glioblastoma (WHO IV).

■ Commentary

The ideal approach to an atrial tumor must provide not only sufficient visualization of the tumor, but also a short path with minimal brain disruption. There are two general options: traversing brain parenchyma (transcortical approaches), or the corpus callosum (interhemispheric approaches). It is important to point out here that the splenium of the corpus callosum does not form the roof of ventricular atrium, and thus, accessing the atrium via a posterior callosotomy requires an *oblique* cut across the corpus callosum (**Fig. 30.19**). A *perpendicular* cut would land the surgeon, not in the ventricle, but in the pineal/superior vermian region instead.

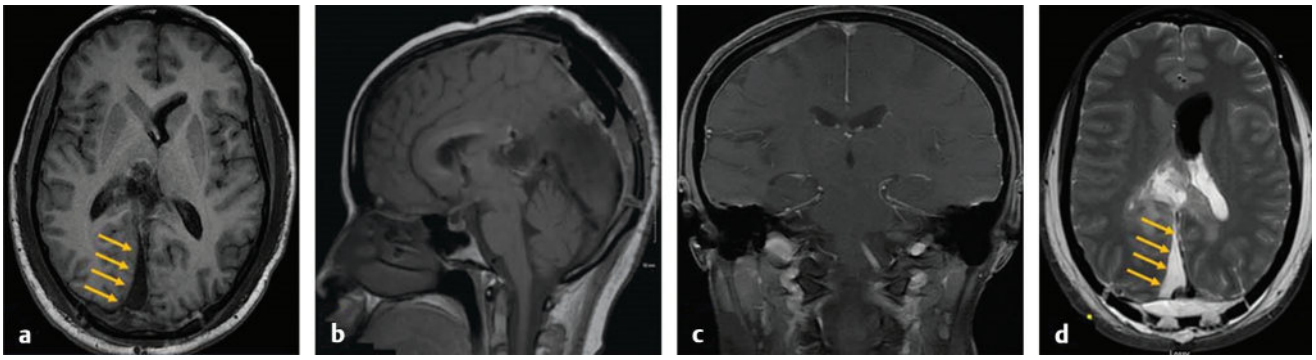


Fig. 30.18 Postoperative MRI images, Case 3. (a–d) Gross total resection and the track of right interhemispheric corridor (*yellow arrows* in **a** and **d**).

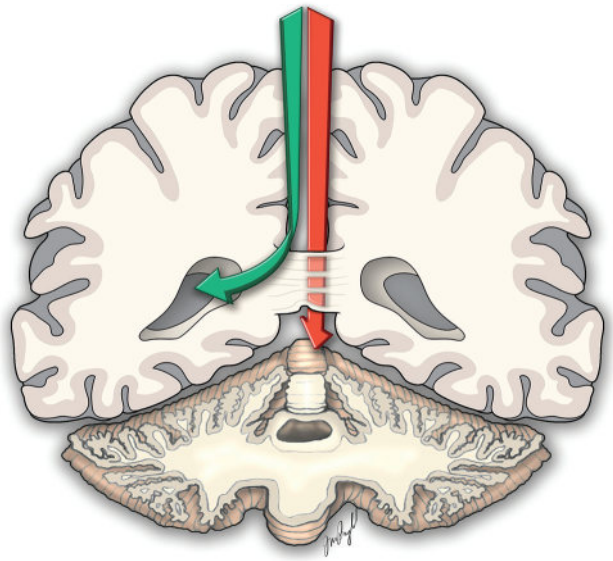


Fig. 30.19 The posterior interhemispheric transcallosal approach. A perpendicular cut (*red arrow*) across the splenium of the corpus callosum would place the surgeon in the pineal region. To access the atrium of the lateral ventricle, an oblique cut (*green arrow*) is used instead.

Multiple factors affect the decision-making process for selection of best surgical corridor and these include the side and size of the lesion, hemispheric dominance, pathology, size of ventricles, neurologic status of the patient, and the surgeon's familiarity with particular approaches (**Fig. 30.20**). For lesions involving the *medial wall* of the atrium and the splenium of the corpus callosum, the interhemispheric posterior transcallosal approach is preferred. Also for *medial wall* lesions, especially ones with extensions into the posterior third ventricle behind the massa intermedia, the ipsilateral interhemispheric parieto-occipital or contralateral transfalcine transprecuneus approach may be appropriate. Tumors located in the *inferior* part of the atrium and posterior parahippocampal gyrus may be suitable for the supracerebellar transtentorial transcollateral sulcus approach. For *inferolateral* lesions, the transtemporal approach utilizes a cortical incision through a portion of the middle or inferior temporal gyrus, and can be adapted to lesions of any size. The disadvantage is risk to visual and language functions. A variant of this is the subtemporal approach, which offers an advantageous lateral route for atrial tumors with feeders from the anterior choroidal artery. It provides direct access to devascularize the tumor while leaving the visual fibers undisturbed. Finally, the parietal transcortical approach (also called the superior parietal lobule approach) is perhaps the one with which neurosurgeons have the most experience. It can access both the medial and lateral walls of the atrium by traversing "minimally" eloquent parenchyma.

Compared to the transprecuneus approach, which was the subject of the first section of this chapter, the lateral transtemporal and superior parietal lobule approaches provide wider exposure to the tumor and allow better control of blood supply, which usually enters inferomedially. Our first two cases (cases 1 and 2) were approached via a posterior temporal transsulcal transcortical approach because it provided the shortest, straightest path to the target. The location and size of cortical incision were chosen based on preoperative diffusion tensor imaging findings, intraoperative surface topography, and data from neuronavigation. Clearly, incision in a sulcus puts one closer to the ependymal surface than one on the nearest gyrus. For case 1, the corticectomy was made at a nadir of a sulcus along the axis of the optic radiation in order to limit injuries (**Fig. 30.12**). In case 2, we exploited a large sulcus to reach the tumor via a trans-sulcal approach through the thinnest cortex (**Fig. 30.13**). Since the tumor compressed the lateral wall of the trigone in both cases, we debulked the tumor early, in order to separate it from the ependymal wall to minimize damage to the optic radiation. It was important to eliminate the vascular pedicle from the anterior and posterior choroidal arteries early to avoid excessive bleeding. Veins from the atrium and occipital horn drain into the ICV and these must be preserved. Although transcortical routes were used, neither of these patients suffered new visual problem after surgery. We believe that strategic placement of the corticectomy and minimal tissue disruption were the keys to this result.

For these two cases, an interhemispheric, transcallosal approach would have involved a much longer distance to target, and for reaching the lateral surface of the tumor, a significant retraction force on the ipsilateral visual cortex would have been required. However, we did use the transcallosal approach in the third case, as it was a direct route to the splenial part of the tumor and gave us bilateral access to tumor components on both sides of the midline (**Fig. 30.16**).

For all the anatomical reasons already discussed in this chapter, entry into the atrium via its medial wall avoids the visual

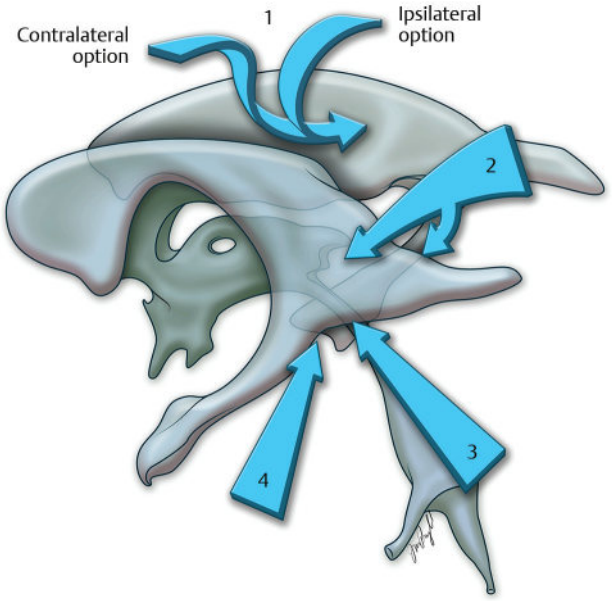


Fig. 30.20 Perspective authors' options to the atrium of the lateral ventricle. 1, for lesions involving the *medial wall* of the atrium and the splenium of the corpus callosum, the interhemispheric posterior transcallosal approach is preferred. Also for *medial wall* lesions, especially ones with extensions into the posterior third ventricle behind the massa intermedia, the ipsilateral interhemispheric parieto-occipital or contralateral transfalcine transprecuneus approach may be appropriate. 2, the parietal transcortical approach (also called the superior parietal lobule approach) can access both the medial and lateral walls of the atrium by traversing "minimally" eloquent parenchyma. 3, tumors in the *inferior* part of the atrium and posterior parahippocampal gyrus may be suitable for the supracerebellar transtentorial transcollateral sulcus approach. 4, for *inferolateral* lesions, the transtemporal approach utilizes a cortical incision through a portion of the middle or inferior temporal gyrus, and can be adapted to lesions of any size. The disadvantage is risk to visual and language functions.

fibers and is theoretically safer. Approaches through the precuneus are thus thought to be preferable, but some counterevidence has emerged. The precuneus has now been shown to play a role in memory, irrespective of cerebral dominance. Tokunaga et al reported the morbidity associated with the transprecuneus approach, such as disturbance of recent memory, thought to be due to either direct damage of the precuneus or manipulation to the crux of the fornix. Furthermore, this approach is limited by the deep and narrow working space and poor visualization of feeding arteries obscured by the tumor. It is also not suitable for large lesions like the ones in our case examples.

In conclusion, even surgery through so-called "safe" areas have risks, and the choice of approach depends on patient factors, tumor characteristics and its relationship with surrounding structures, and finally, surgeon's experience. We believe surgeons dealing with these complex tumors should be versatile performing every approach, so that all options are available for the patient.

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31 Third Ventricle

Cristian Gragnaniello and Walter C. Jean

Keywords: transchoroidal, interforniceal, internal cerebral vein, colloid cyst, craniopharyngioma, germ cell tumor

■ Case Presentation

A 40-year-old African American man with a history of recent myocardial infarction presented to the emergency department after he was woken up by a severe headache that did not respond to nonsteroidal anti-inflammatory drugs. At presentation to our hospital, he explained that similar episodes of headache had occurred in the last few months, but this particular episode was accompanied by blurry vision as well. On examination, he was neurologically intact and had no evidence of papilledema. A head computed tomography (CT) done on arrival showed significant hydrocephalus, and a large, hyperdense mass in the third ventricle. The brain magnetic resonance imaging (MRI) obtained subsequently demonstrated an anterior third ventricular cystic lesion. Its size, measured at 36 mm in diameter, was sufficient to stretch the fornices in a superior direction. It appeared to be blocking the foramen of Monro, and was suspicious as the cause of the hydrocephalus. The lesion was dark on T1- and bright on T2-weighted images, with no enhancement on gadolinium administration (**Fig. 31.1**).

Questions

1. What is the differential diagnosis of the imaging findings?
2. How does the differential diagnosis influence the next step in management?
3. If a surgical approach is planned, what are the most important considerations for a successful outcome?

■ Diagnosis and Assessment

A variety of neoplastic lesions can present in this exact location in the third ventricle. However, the vast majority of them, including craniopharyngioma, giant pituitary adenoma, ependymoma, and central neurocytoma, enhance with gadolinium in various patterns, and can therefore be eliminated from our consideration. In contrast, a subependymoma does not enhance with gadolinium, but this tumor usually presents in the lateral ventricles.

As rare as they are, vascular lesions need to be considered, as poor surgical preparation for them can lead to catastrophic consequences. A giant thrombosed aneurysm at the basilar apex can present as a third ventricular mass, but one would expect surrounding flow voids and a heterogeneous signal within the mass. Finally, neurocysticercosis can be considered, but our patient's demographic characteristics, the lack of calcification, the solitary presence and size of his lesion, all point away from this diagnosis.

The size, shape, and MRI signal characteristics of the lesion in our 40-year-old patient all point to the diagnosis of a colloid cyst. The vast majority of these lesions are detected in patients without symptoms, and the cyst smaller than 10 mm. The discussion of whether these patients require treatment is presented in Chapter 29. There are few neurosurgeons who would protest against treating a patient with a 36-mm colloid cyst presenting with significant hydrocephalus. As such, an external ventricular drain (EVD) was placed in the intensive care unit moments after his arrival on the unit. Fortunately, a solitary right-sided EVD was enough to control his hydrocephalus, indicating that there was still some cerebrospinal fluid (CSF) flow around the foramen of Monro.

■ Anatomical and Therapeutic Considerations

Although this colloid cyst was positioned in the typical location in zone I of the third ventricle, between the lamina terminalis and the massa intermedia, it did have some unusual anatomic features. It was unusually large, compared to most colloid cyst

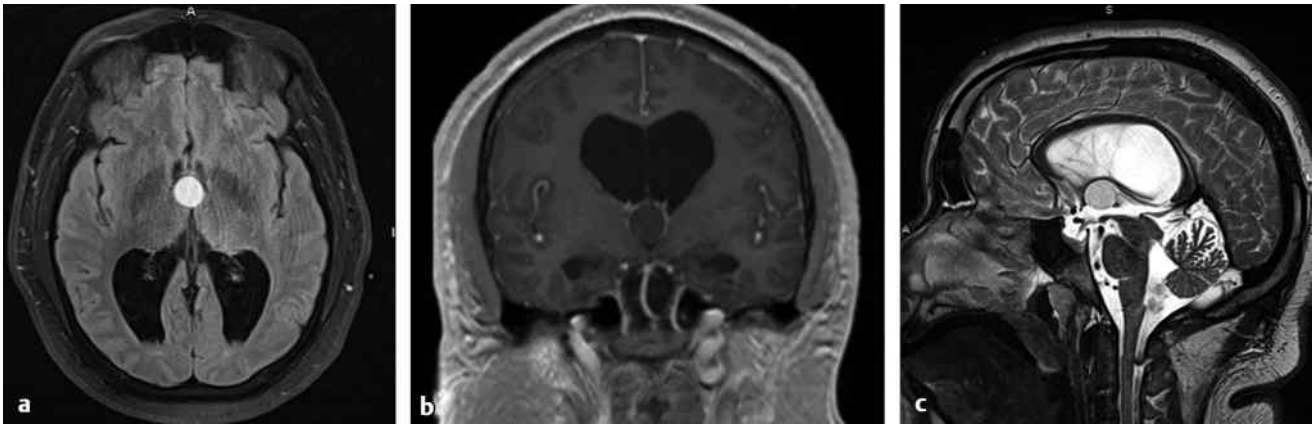


Fig. 31.1 Preoperative MRI. **(a)** FLAIR axial, **(b)** T1-weighted coronal image with gadolinium, and **(c)** T2-weighted sagittal image showing a cystic lesion in the anterior third ventricle causing hydrocephalus. The lesion was dark on T1, bright on T2 and showed no enhancement with gadolinium.

at presentation, and almost the entire mass was *behind* the foramen of Monro. This aspect posed some challenges for its surgical removal. In a healthy, young man, the surgical goal must be total removal of the mass. Temporizing measures, such as stereotactic cyst aspiration, would make very little sense in this patient, as reaccumulation and further enlargement of the cyst would lead to symptoms worse than hydrocephalus, even if this problem were addressed by ventriculoperitoneal shunting.

Options for Approach

In recent years, advancements in equipment and skills have made neuroendoscopy an attractive technique for removal of colloid cysts. In turn, the paucity of blood supply and location at the foramen of Monro make most colloid cysts enticing targets for this "minimally invasive" technique. However, clinical results still show that endoscopic techniques are inferior to open microsurgery in rates of recurrence and reoperation. The endoscopic technique is dependent on transformaninal deflations of the cyst and detachment of the cyst wall from the roof of the third ventricle. The location of our patient's cyst behind the foramen and its large size made it ill-suited for this technique.

There are various well-described techniques to access the anterior third ventricle by open microsurgery (**Fig. 31.2**). Although it is one of the oldest methods, the translaminar terminalis route, whether by Dandy's original description, or the modern day, subcallosal variant, is rarely utilized. As such, there is insufficient clinical data to know whether the functional outcome for the patients is better or worse when compared to more popular approaches.

Far more clinical data are available for approaches which access the third ventricle from a superior direction utilizing either the foramen of Monro or the choroidal fissure to reach the target. The transcortical–transforaminal approach is applicable to patients with hydrocephalus with an expanded foramen. Widely taught to neurosurgical trainees as risky for postoperative seizures, this approach has been vindicated in recent data which refute this myth. In reality, the biggest disadvantage of this approach is perhaps the unilaterality of access, since the surgical corridor is almost uniformly through the right frontal lobe, and thus large lesions, with significant left-sided components, may pose serious challenges.

Alternatively, instead of proceeding through the frontal cortex, the corridor through the interhemispheric fissure and the corpus callosum can provide excellent access into the ventricular system. The dissection of the interhemispheric fissures must be done carefully in order to preserve the draining veins into the superior sagittal sinus (SSS) and the pericallosal arteries, but once past these, the transcallosal approach offers several options to reach the anterior third ventricle. Of these, the route through the choroidal fissure is perhaps the most commonly utilized in modern neurosurgery. This fissure is the natural cleft between the fornix and the thalamus. It contains the choroid plexus, which in turn is anchored to the fornix by the tenia fornices and to the thalamus by the tenia thalami. The contents of the fissure are functionally inert, and the natural cleft therefore provides a safe way to open the roof of the third ventricle for surgical access and maneuvering (**Fig. 31.3**).

In the transchoroidal approach, the choroidal fissure can be dissected on either side of the choroid plexus. On the thalamus side, it would be the tenia thalami that is traversed, whereas on the fornix side, the tenia fornices. Here, once again, there is no data supporting which route is better. Most surgeons prefer the tenia fornices, as dissecting here makes it less likely that the important thalamostriate vein would be stretched and

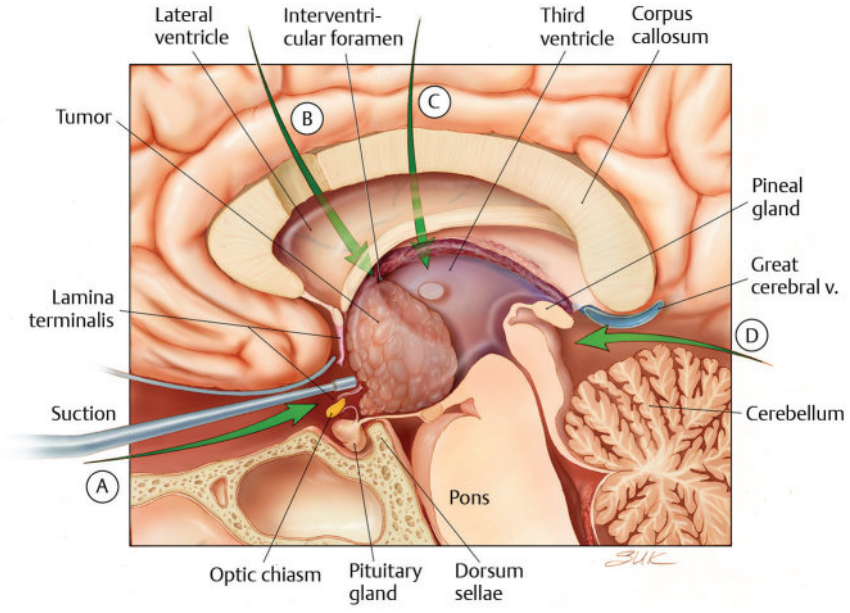


Fig. 31.2 Microsurgical approaches to the third ventricle. (A) Anterior–inferior approaches are primarily through the lamina terminalis. The approach to the lamina terminalis can be either via a unilateral or bilateral subfrontal approach. Superior approaches can be **(B)** anterior or **(C)** posterior. These approaches lead the surgeon into the lateral ventricle. Once within the lateral ventricle, the third ventricle can be entered either through **(B)** the foramen of Monro or **(C)** a transchoroidal corridor. **(D)** For posterior third ventricle tumors that occupy the pineal region, posterior approaches can be used, including the supracerebellar infratentorial approach or the occipital transtentorial approach. (Reproduced from Badie B, ed. Neurosurgical Operative Atlas. Neuro-Oncology. 2nd ed. Thieme; 2006.)

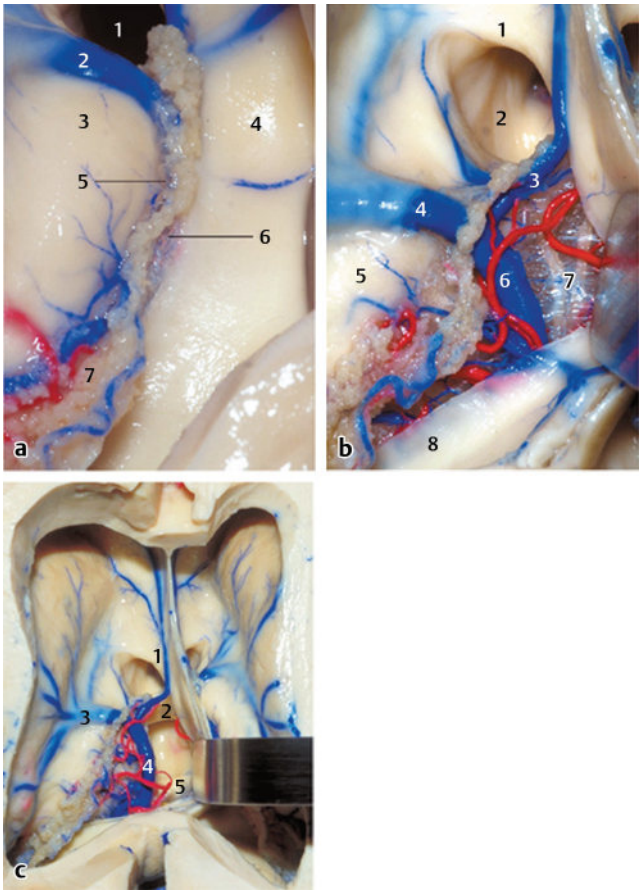


Fig. 31.3 Microsurgical anatomy of the choroid fissure and the transchoroidal approach. (a) Magnified view of the choroid plexus in the body of the left lateral ventricle. The choroid plexus is attached medially to the body of the fornix by the taenia fornicis and laterally to the thalamus by the taenia choroidea. The taenia is actually a continuation of the ependyma that covers the ventricular cavity over the choroid plexus of the lateral ventricle. 1, foramen of Monro; 2, thalamostriate vein; 3, thalamus; 4, fornix; 5, taenia choroidea; 6, taenia fornicis; 7, choroid plexus and branches of the medial posterior choroidal artery. **(b)** Superior view. After splitting the taenia fornicis and the superior membrane of the tela choroidea, the internal cerebral vein, the branches of the medial posterior choroidal artery, the inferior membrane of the tela choroidea, and the choroid plexus of the third ventricle come into view. 1, column of the fornix; 2, foramen of Monro; 3, anterior septal vein; 4, thalamostriate vein; 5, thalamus; 6, internal cerebral vein; 7, inferior membrane of the tela choroidea and the choroid plexus of the third ventricle; 8, crus of the fornix. **(c)** Superior view. After opening the inferior membrane of the tela choroidea, the cavity of the third ventricle, from the foramen of Monro to the posterior commissure is reached. When necessary, the anterior septal vein can be sacrificed to gain additional working space. 1, anterior septal vein and column of the fornix; 2, massa intermedia; 3, thalamostriate vein; 4, internal cerebral vein; 5, posterior commissure. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

injured (**Fig. 31.4**). Proponents of this technique believe that the septal vein can be sacrificed if necessary. Other surgeon would argue for using the tenia thalami for opening the choroidal fissure, as this is less likely to damage the fornix, and in this technique, the choroid plexus would in fact serve as a sort of cushion to protect the ipsilateral fornix (**Fig. 31.5**). With this equivocation, it is the senior author's pragmatic approach that whichever side of the choroid plexus offers the most expedient opening in any given patient, that is the side to use. An aperture of about 2 cm through the choroidal fissure, whichever side of the choroid plexus one takes, provides a wide view of the anterior half of the third ventricle.

Finally, the transcallosal route offers another, more medial approach into the third ventricle, and that is through the septum pellucidum and between the two columns of the fornix. This so-called interforniceal approach has been shown to offer a wider opening into the target ventricle than the transchoroidal route (**Fig. 31.6**). Although transchoroidal surgery puts the ipsilateral fornix in harm's way, the interforniceal technique involves both fornices. Clinical data is, again, inadequate to tell us whether the theoretical fear of bilateral forniceal injury in fact translates into poorer clinical outcome for the patients' memory functions.

Questions

1. What are the major risks during dissection of the interhemispheric fissure prior to reaching the corpus callosum?
2. For our patient's lesion, which lateral ventricle should be "opened" after traversing the corpus callosum?
3. How long a stretch of the corpus callosum would you open for such a transcallosal approach?

Approach of Choice

Among the choices of the most commonly utilized approaches to the third ventricle, we chose to use the transcallosal transchoroidal route. Given the size of the lesion, and its position behind the foramen of Monro, we did not think that a transforaminal resection was feasible. In contrast, the transcallosal transchoroidal approach would provide a wide, near-midline access to both sides of the lesion and enough exposure to the posterior pole for an efficient cyst removal.

■ Description of the Technique

The Three Approach Elements

Corridor: interhemispheric

Craniotomy: frontal

Modifier: corpus callosotomy, dissection of choroidal fissure

After establishing general endotracheal anesthesia, the patient was placed supine with the head and neck in neutral position for the transcallosal transchoroidal approach. The incision we could make was restricted by the previous one used for his EVD.

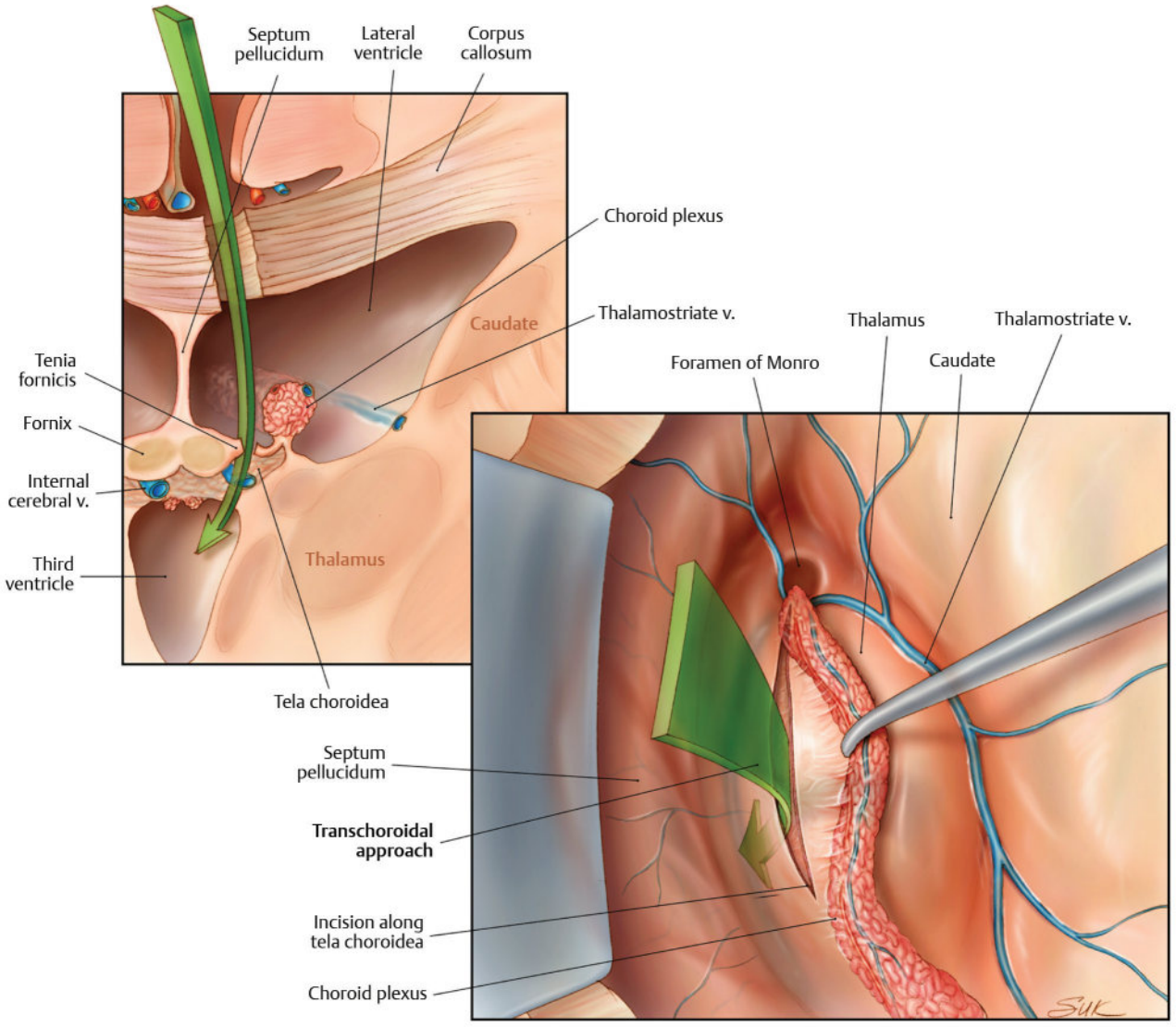


Fig. 31.4 Illustration of the transchoroidal approach using the tenia fornicis. Upper left shows a coronal view of interhemispheric transcallosal entry into lateral ventricle. Passage into the third ventricle is made on the medial side of the choroid plexus (along the tenia fornicis). (Lower right) An incision is made along the tenia fornicis through the tela choroidea. The surgeon can then dissect between both internal cerebral veins to be free of any vessels. (Reproduced from Badie B, ed. Neurosurgical Operative Atlas. Neuro-Oncology. 2nd ed. Thieme; 2006.)

Had we chosen the side contralateral to the EVD, we might have used a straight incision parallel with the coronal suture, or a "U-shaped" incision based laterally. For our patient, we chose to incorporate the 2-cm straight incision used for his EVD into a new "lazy S-shaped" incision to expose the right frontal area, from the midline to 5 cm lateral on the coronal suture (see **The Three Approach Elements**).

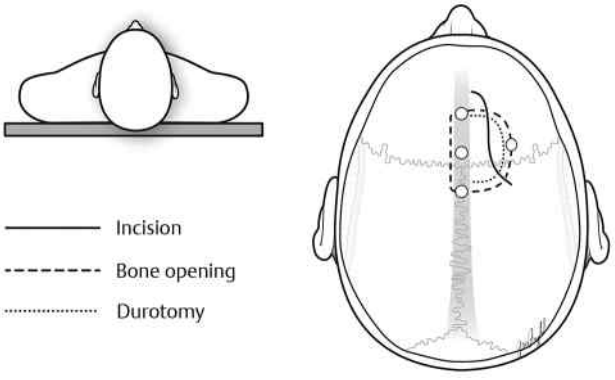
Operative Setup

Position: supine, head neutral

Incision: "lazy S" incorporating incision at Kocher's point

Bone Opening: 5 cm × 3 cm right frontal exposing SSS

Durotomy: "U-shaped" based on SSS



Three burr holes were made, one at the junction of the coronal and sagittal sutures, and then one anterior and one posterior

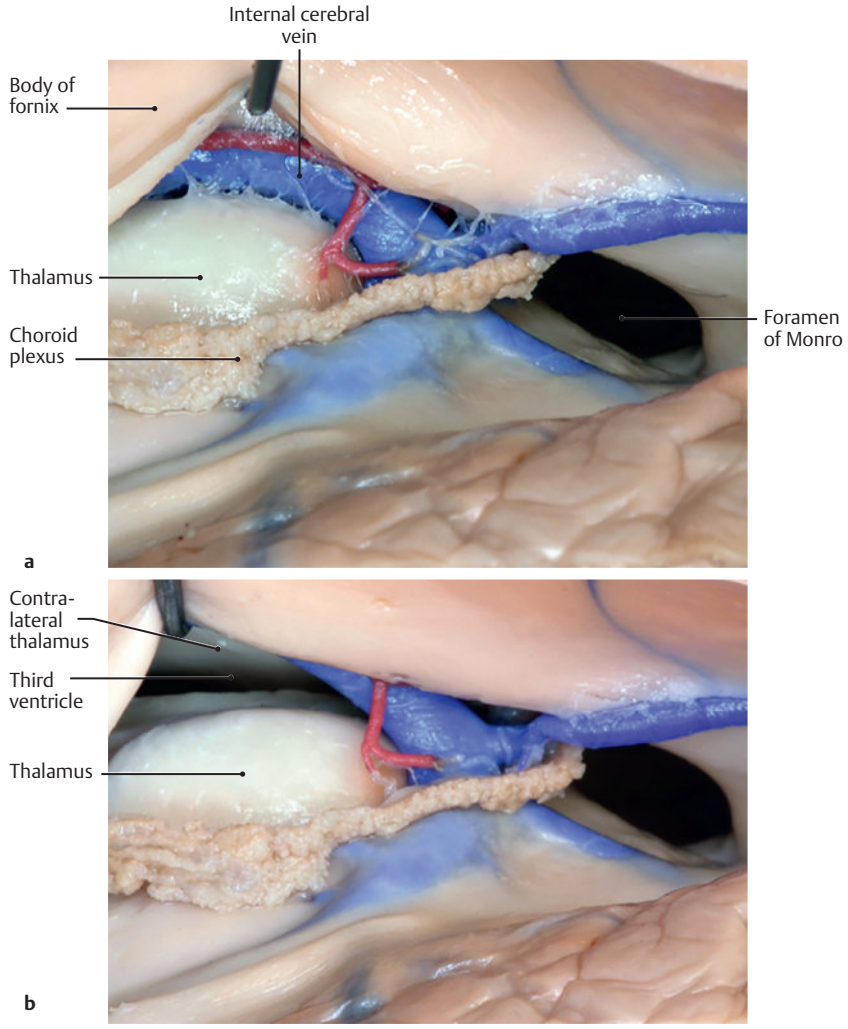


Fig. 31.5 Transchoroidal approach in the lateral position using the tenia thalami. **(a)** View of the lateral ventricle with the patient in a lateral position and the relation among the thalamus, fornix, and choroid plexus. The choroidal fissure can be opened between the fornix and the choroid plexus or between the thalamus and the choroid plexus. **(b)** Opening the fissure on the thalamic side minimizes possible injury to the fornix. The choroid plexus is used as a cushion to minimize forniceal retraction while opening the choroid fissure on the thalamic side. (Reproduced from Spetzler R et al. Color Atlas of Brainstem Surgery. 1st ed. Thieme; 2017.)

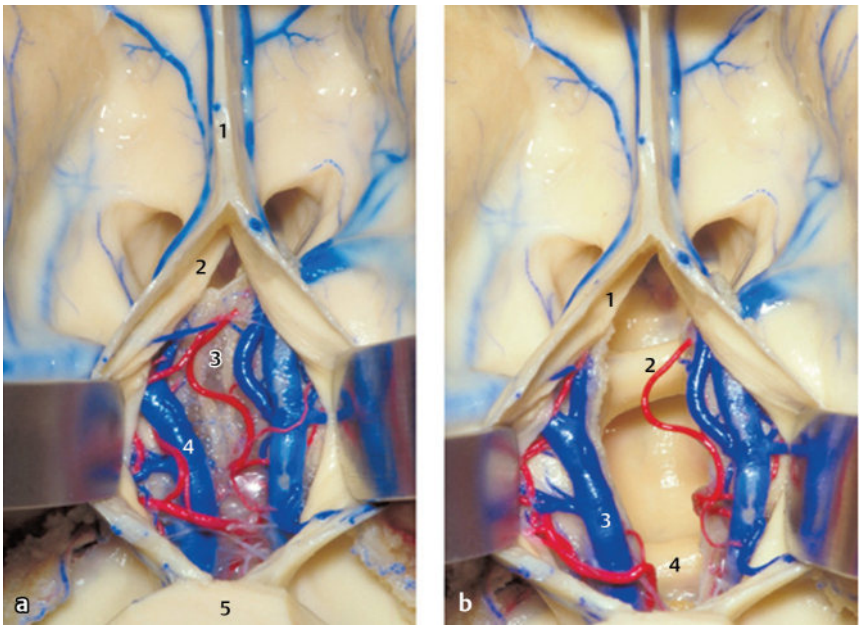


Fig. 31.6 The interforniceal approach. **(a)** Superior view. After entering the body of the lateral ventricle, the dissection continues between the two leafs of the septum pellucidum down to their attachment at the body of the fornix, then the body of the fornix is divided in the midline. 1, septum pellucidum; 2, fornix; 3, inferior membrane of the tela choroidea; 4, internal cerebral vein; 5, splenium of the corpus callosum. **(b)** After the opening of the inferior membrane of the tela choroidea and the choroid plexus of the third ventricle, the cavity of the third ventricle is seen. 1, fornix; 2, massa intermedia; 3, internal cerebral vein; 4, posterior commissure. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

to this. The middle burr hole would be used purely to dissect the bone off the SSS. A final burr hole was placed laterally, and the crantiome was then used to make a 5 × 3 cm bone flap, with the midline cut just to the left of the SSS, skipping the middle burr hole. Some surgeons would consider exposing the SSS unnecessary risky for this procedure, but in our experience, it

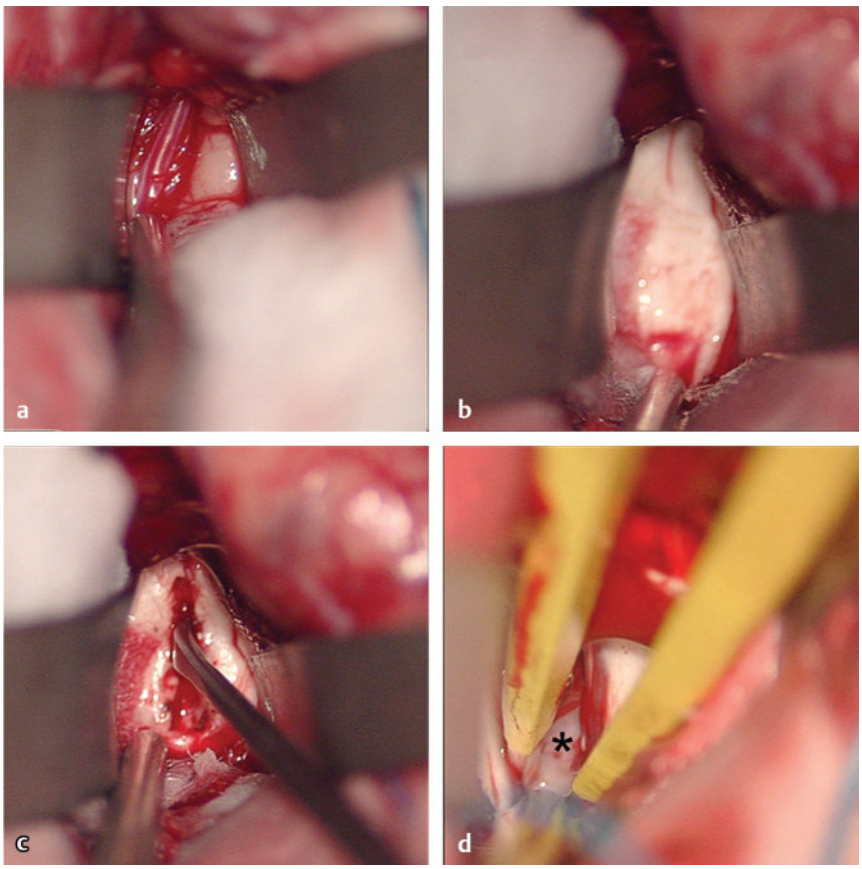


Fig. 31.7 Intraoperative views of the transcallosal transchoroidal approach. **(a)** The right pericallosal artery was seen after dissection of the cingulate fissure; **(b)** the corpus callosum appeared as a bright white structure at the end of the interhemispheric fissure; **(c)** a 2-cm stretch of the corpus callosum was opened to access the lateral ventricle; **(d)** the choroidal fissure was opened to reveal the lesion in the third ventricle (*).

allows just a bit more freedom to place a retractor on the falx cerebri to widen the corridor (see **Operative Setup**).

The dura was opened as a "U" based medially, and an area near the midline was found devoid of draining veins into the SSS. Using this area, dissection was taken deep into the interhemispheric fissure and as mentioned above, the falx was retracted to the left to assist this maneuver. The plane between the cingulate gyri is often quite adherent, but in this patient, it dissected quite easily apart. With that, both pericallosal arteries were seen and protected (**Fig. 31.7a**). A 2-cm stretch of the corpus callosum was opened in an anterior–posterior direction, and we landed in the left lateral ventricle (**Fig. 31.7c**).

As expected, hardly any part of the colloid cyst was visible through the foramen of Monro. The thalamostriate and septal veins were identified, as was the choroid plexus posterior to them. Using a micro-dissector, this was examined carefully and there seemed to be a clearer plane on the tenia thalami side. This was opened with bipolar forceps, used in a spreading motion (**Fig. 31.7d**). When necessary, bipolar cautery was used at low amplitude to stop any bleeding in the fissure. In this manner, about 2 cm of the fissure was opened, giving us a clear view of the colloid cyst in the third ventricle.

The cyst was punctuated, and nearly all the content was evacuated directly into the suction. The cyst wall was then mobilized into the left lateral ventricle, and a cottonoid was placed into the space it previously occupied in the third ventricle to prevent it from prolapsing back into it. The wall was then dissected away from the roof of the third ventricle, being especially careful not to harm the fornix on either side. Since the left fornix was in full view, naturally attention was particularly focused here. In this way, the colloid cyst was completely removed.

A ventricular drain was once again left in the lateral ventricle, placed under direct vision. The dura was closed and the bone flap replaced. The rest of the closure was done in standard fashion.

Surgical Pearls

1. Some surgeons advocate using the lateral position for this operation, putting the ipsilateral side down. This way, gravity can be used in place of retractors to mobilize the ipsilateral frontal lobe away from the interhemispheric fissure. Had our patient's body habitus been smaller, we would have considered using this technique, but as he was quite large, it was simpler and perhaps safer to keep him supine and neutral.
2. In the interhemispheric fissure just deep to the inferior sagittal sinus, the two cingulate gyri are often stuck to each other quite firmly. Compared to the sylvian fissure, this is harder to "open." Knowing this and exercising patience is critical, as the pericallosal arteries lurk nearby and must be preserved and protected. Injury to these can cause leg weakness, and this is one of the major risks of any interhemispheric approach.
3. Since our patient's lesion is positioned perfectly in the midline, whether the transcallosal route landed us to the right or left lateral ventricle made no difference. This laterality issue may be important for other lesions that are asymmetric across the midline.

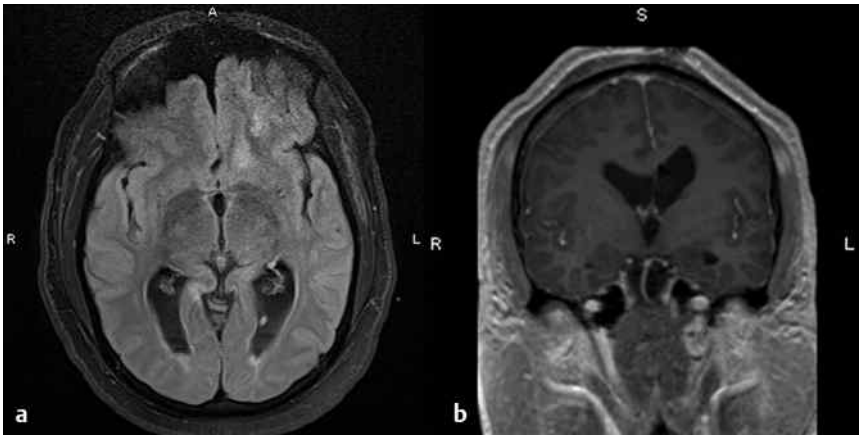


Fig. 31.8 Postoperative MRI. (a) FLAIR axial and **(b)** T1 coronal views showing complete removal of the colloid cyst.

■ Aftercare

The patient awoke from surgery much as he was before going into it. Our practice is to leave the EVD for 5 days for removal of surgical debris, and with this, hopefully avoid clogging the arachnoid granulation and subsequent hydrocephalus. His memory function was a bit worse initially after the operation, but within a week, he was able to name all 11 of his children and their ages. Unfortunately, despite aggressive CSF/debris drainage, and having emptied his third ventricle of the colloid cyst (**Fig. 31.8**), he was dependent on the EVD for intracranial pressure control. Near the end of the second postoperative week, a ventriculoperitoneal shunt was placed, and with that, he was discharged with rehabilitation.

On follow-up several months after the operation, he complained that his memory function was impaired enough that he was unable to resume his previous employment. Luckily for his children, he was still able to remember all of them. He also complained of left leg weakness, but this was undetectable on physical examination. His shunt continued to function without problems.

■ Possible Complications and Associated Management

As with many skull base operations, there are approach-related and resection-related risks to this operation. Some of the nuances of the approach have already been mentioned. The bone opening should be longer (anteroposterior dimension) than it is wide (L/R dimension). The reason for this is to provide more options for where to dissect the interhemispheric fissure. The goal is to stay away from draining veins, and mitigate the chances of either harming these, or needing to sacrifice them to gain access to the fissure.

Certainly, the ipsilateral pericallosal artery is placed under the highest risk during the transcallosal approach, as frequently, pressure is placed on this vessel to gain adequate exposure into the lateral ventricle. To prevent strangulation or spasm in this vessel during the operation, it must be well-protected once it is visualized. The rest of the operation is deep to it, and the surgeon's attention would be focused there for the vast majority of the procedure. If exposed, any movement in or out of the operative field can damage this artery.

Once into the lateral ventricle, complication avoidance is mostly about the fornices. There is naturally a risk to this when opening the choroidal fissure. Some surgeons prefer to dissect the fissure on the tenia thalami side, which allows the choroid plexus to act as a "cushion" for the fornix during the resection. Colloid cysts are adherent to the roof of the third ventricle, of which the fornices are part. Large cysts may be adherent to longer stretches, and meticulous microsurgical technique is the best way to avoid damage to them.

Despite the utmost care in hemostasis and avoiding slippage of the cyst contents into the ventricular system, surgical debris inevitably accumulate after surgery within the ventricular system. It is therefore our routine practice to drain the CSF aggressively for 5 days or until the CSF significantly clears. We believe that this lowers the shunt rate after these procedures, but unfortunately for this patient, he still required shunt placement despite this management.

Perspective

Cody L. Nesvick and David J. Daniels

■ Introduction

In the preceding section, the interhemispheric transcallosal transchoroidal approach was discussed in detail. Overall, we agree with its application for the colloid cyst in the third ventricle, as well as with the decision to utilize an open versus an endoscopic approach. The decision to use any particular approach to the third ventricle is highly dependent on details of each patient and tumor, but surgeon preference and experience also play important roles. In this section, we describe our perspective on surgery of the third ventricle with an emphasis on the interforniceal approach.

■ Anatomical and Therapeutic Considerations

There are two major considerations for planning surgery on third ventricular lesions: the "angle of attack" and the corridor for accessing the third ventricle. An optimal approach must offer an adequate view of the local venous anatomy, and because the internal cerebral veins (ICVs) reside in the roof of the third ventricle, dissection around these delicate vessels is unavoidable for any approach from a superior trajectory.

Angle of Attack

In general, a transcortical approach offers an oblique approach to the third ventricle via the foramen of Monro. It is most appropriate for small lesions that are eccentric to one side and/or protrude through the foramen. For such lesions, like most colloid cysts, the transcortical approach provides an operative trajectory along the long axis of the lesion, minimizing the need for additional retraction. However, unless there is massive hydrocephalus, the surgical corridor can be limited by the location of the initial corticectomy, and suboptimal placement of the corticectomy will force the surgeon to "create space" with excessive retraction.

In comparison, the interhemispheric transcallosal approach offers a straight midline approach with much broader exposure and direct access to the third ventricle. However, it exposes significantly more vascular structures than the transcortical approach, and the surgeon must be cautious in preserving these structures throughout the dissection. Bridging veins to the sagittal sinus are often encountered during the initial approach. To minimize the need to sacrifice important bridging veins, and for the purpose of defining deep venous anatomy, we routinely obtain a preoperative MR venography to plan our durotomy and operative corridor. Additionally, the pericallosal and callosomarginal arteries must be visualized and protected during the approach to avoid lower extremity paresis.

Access to Third Ventricle

Once the 'angle of attack" is defined, the surgeon may choose from one of three corridors to access the third ventricle: transforaminal, transchoroidal, or interforniceal. The transforaminal approach utilizes the foramen of Monro as a natural corridor into the third ventricle and is often the best approach for smaller lesions that arise near the foramen. It is also suitable for large tumors that significantly expanded the foramen, especially when there is associated hydrocephalus which naturally enlarged the surrounding surgical space.

When the foramen of Monro is normal in size, large lesions of the third ventricle require either a transchoroidal or interforniceal approach for access. The nuanced difference between these two is explored with the next two case examples.

■ Case Presentation 1

A 12-year-old boy with a history of trisomy 21 and associated cardiac cushion defect presented with diabetes insipidus. His brain MRI showed a diffusely contrast-enhancing lesion filling the sella and suprasellar space, extending into the third ventricle. On coronal images, it was noted that his foramen of Monro was slighted expanded, and his lateral ventricles were also slightly larger than normal (**Fig. 31.9**).

For this patient, we opted for the interhemispheric transcallosal transchoroidal approach, using the technique much like that already described in the first section of this chapter. The anterior septal vein was cauterized and divided to facilitate expansion of the choroidal fissure. A gross total resection was achieved, and a right frontal EVD was left in place. Histopathological examination revealed a craniopharyngioma. In the immediate postoperative period, he developed central hypocortisolism and hypothyroidism, which were promptly recognized and treated. His vision remained intact, and his EVD was removed without problems.

■ Case Presentation 2

An 18-year-old man presented with headache. A CT scan was notable for a hypodense mass in the middle and posterior aspect of the third ventricle associated with lateral ventriculomegaly. A brain MRI was performed which demonstrated a multiloculated, mixed-intensity cystic lesion with peripheral contrast enhancement (**Fig. 31.10**). He initially underwent a combined endoscopic third ventriculostomy and biopsy, which was diagnostic of a nongerminomatous germ cell tumor. He was initiated on chemotherapy according to the Children's Oncology Group protocol, but his tumor demonstrated 30% growth midway through chemotherapy. As such, surgical resection was recommended.

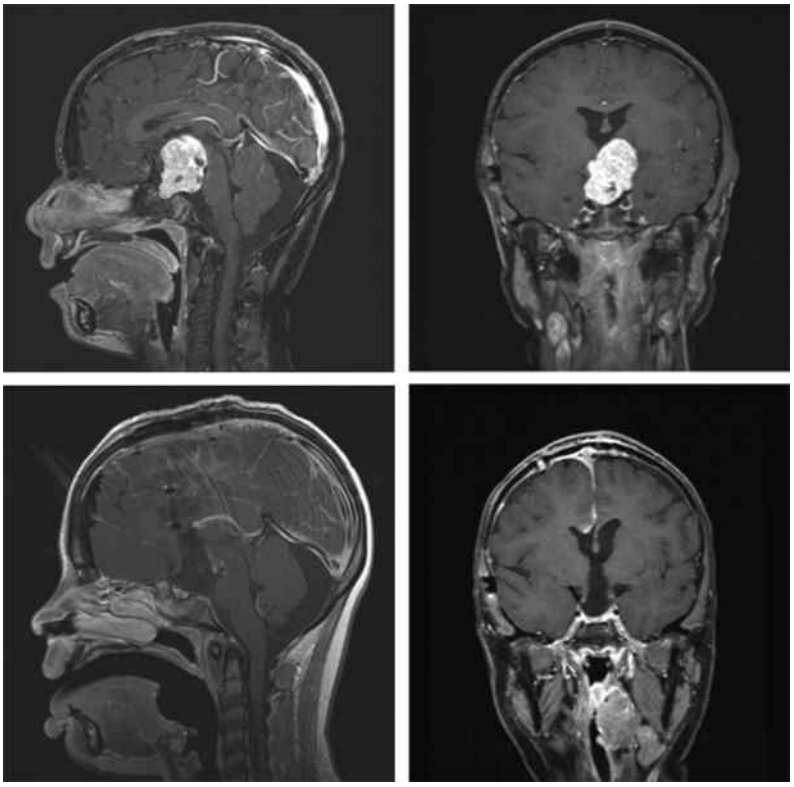


Fig. 31.9 Case 1: Transchoroidal approach. Preoperative (*top row*) and postoperative (*bottom row*) contrast-enhanced, T1-weighted MRIs demonstrating a uniformly enhancing lesion filling the sellar and suprasellar spaces and extending into the third ventricle. The right foramen of Monro was enlarged, and no cavum septum pellucidum was present. A gross total resection was achieved using an interhemispheric transchoroidal approach.

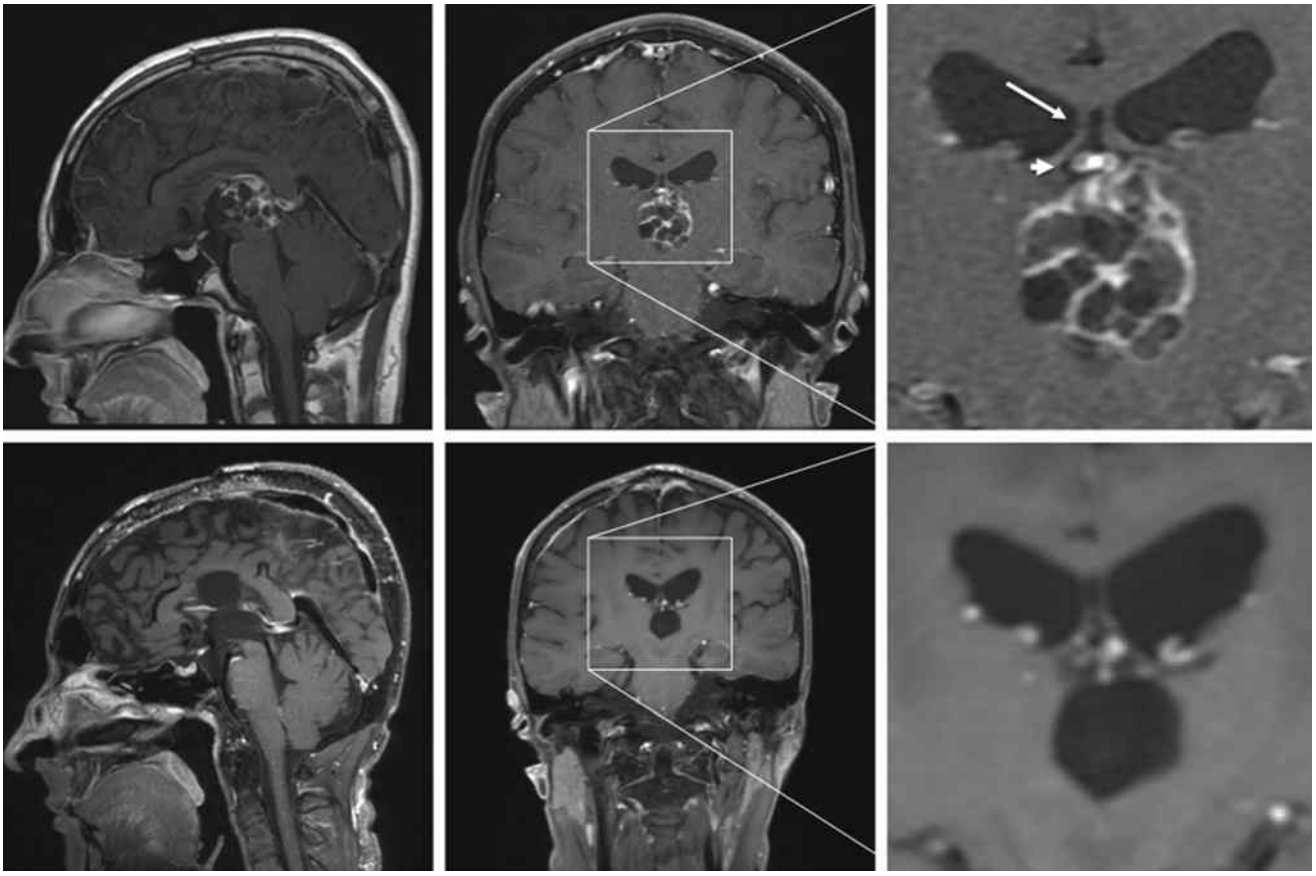


Fig. 31.10 Case 2: Interforniceal approach. Preoperative (*top row*) and postoperative (*bottom row*) MRIs demonstrating a contrast-enhancing, multiloculated cystic lesion in the posterior third ventricle. A cavum septum pellucidum (*long arrow*) was present, and the internal cerebral veins (*short arrow*) were seen directly overlying the tumor in the midline. The tumor was resected en bloc using an interhemispheric transcallosal interforniceal approach.

The large tumor was located in the middle-to-posterior thirds of the third ventricle. Importantly, a cavum septum pellucidum (CSP) was present. Because of these features, an interhemispheric transcallosal interforniceal approach was chosen.

■ Description of the Technique

The patient was positioned supine with the neck flexed to facilitate anterior visualization while maintaining adequate venous outflow through the neck. We used a lazy S-shaped incision approximately 1 cm behind and roughly parallel to the coronal suture. This keeps the craniotomy and approach well anterior of the precentral gyrus. A right-sided approach was used with two burr holes placed along the SSS and one on the right side on the coronal suture. A square-shaped craniotomy was elevated and the dura was opened in a U-shape, based medially. The rest of the procedure was performed under the operating microscope.

We generally utilize dynamic retraction, and therefore operate with the microscope eyepieces 180 degrees apart, allowing a surgical assistant to help with hand-held retractors. Once under the microscope, the falx cerebri was followed to its termination, and any local arachnoid adhesions were dissected away using micro-scissors and insulated bipolar tips. The pericallosal arteries were then visualized and gently displaced laterally to facilitate further dissection. A 2-cm callosotomy was then performed with bipolar and suction at true midline as determined by intraoperative navigation. In this patient, it was crucial to enter the ventricular system *between* the leaves of the septum pellucidum and into the CSP.

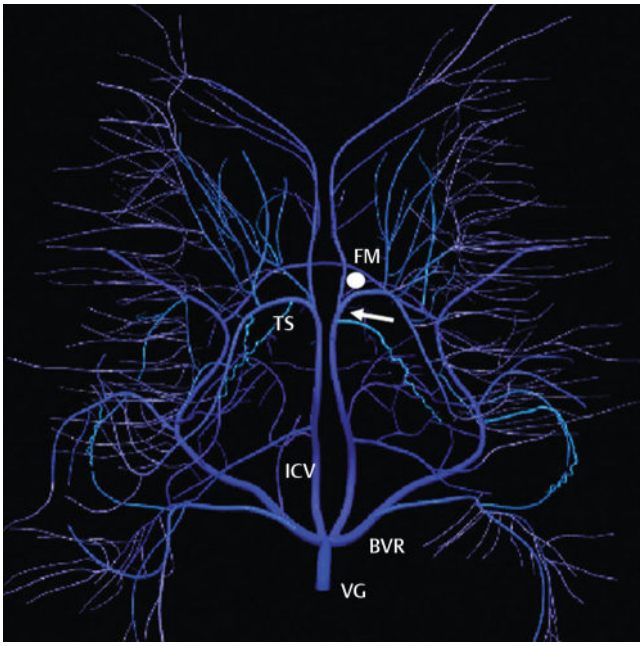


Fig. 31.11 The venous angle. The U-shaped junction between the thalamostriate (TS) and internal cerebral vein (ICV) is called the venous angle (*arrow*). The usual location of this is at the posterior margin of the foramen of Monro (FM). When this joining occurs posterior to this usual location, it is called a false venous angle. When this is found, the affected ventricle should be used for initiation of the transchoroidal approach, as the choroidal fissure may open without sacrifice of the anterior septal vein. BVR, basal vein of Rosenthal; VG, vein of Galen. (Reproduced from Runge V, ed. Imaging of Cerebrovascular Disease. 1st ed. Thieme; 2016.)

With further dissection through his CSP, the third ventricle was entered. On passing through the roof of the third ventricle, small choroidal vessels were encountered. The fornices and ICVs were lateral to the operative field at this point, and the tela choroidea was opened for a wide view of the third ventricle and tumor. The tumor was resected en bloc without notable events.

■ Aftercare

Postoperatively, the patient suffered from akinetic mutism and anterograde amnesia. The former resolved within two

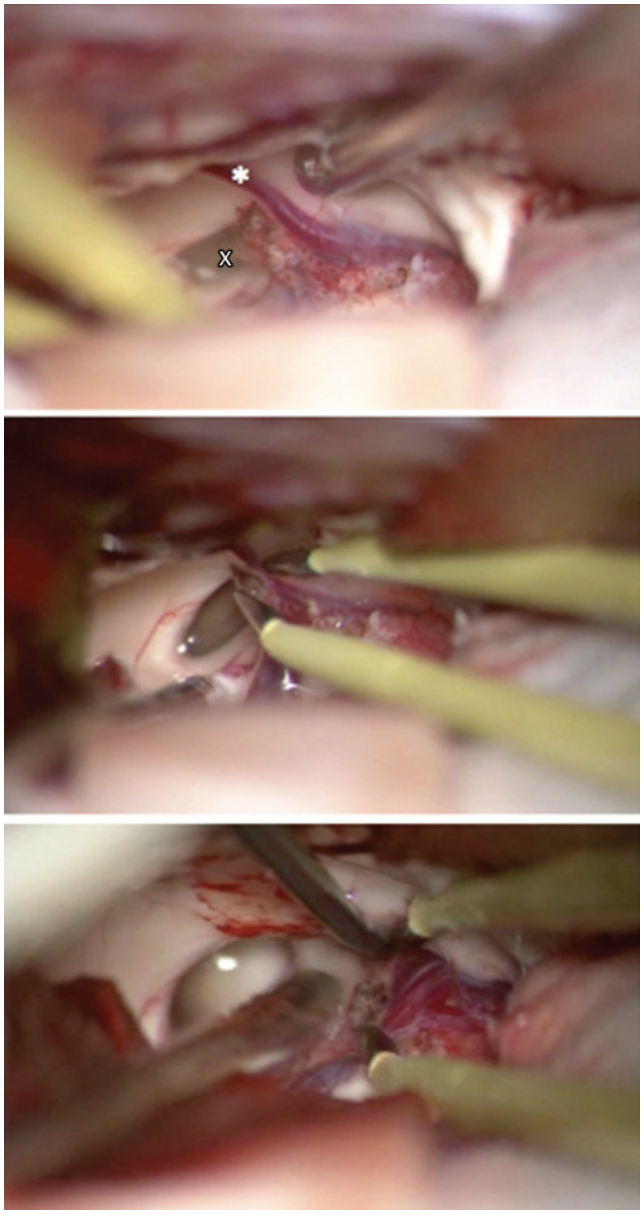


Fig. 31.12 Anterior septal vein, Case 1. On accessing the left lateral ventricle, the septal vein (*), thalamostriate vein and choroid plexus were visible at the foramen of Monro (X) (*top*). To facilitate exposure of the third ventricle, the septal vein was coagulated and divided, allowing for exposure of the venous angle (*middle*). The choroidal fissure was then opened, the fornix was gently displaced medially and the internal cerebral vein was moved laterally along with the choroid plexus to fully visualize the third ventricle (*bottom*).

weeks after surgery, and the latter followed in the next several months. He has since graduated from high school and is a productive college student.

■ Commentary

Transchoroidal Route

As the subject of the first section of this chapter, much has been said about this option. We have only to add that the choice of which lateral ventricle to use for opening the choroidal fissure sometimes matters for this off-midline approach.

The characteristic U-shaped junction of the thalamostriate vein and the ICV adjacent to the posterior margin of the foramen of Monro is referred to as the venous angle (**Fig. 31.11**). When the junction lies *behind* the posterior margin of the foramen of Monro, this variation is called the *false* venous angle. If this is found on preoperative studies of the venous anatomy, then the side on which it is discovered should be used for the transchoroidal dissection, since the false venous angle allows some opening of the choroidal fissure without sacrificing the anterior septal vein, as was needed in our first case example (**Fig. 31.12**).

Interforniceal Route

In our second case example, a CSP was present, and offered a natural, wide, straight midline corridor between the fornices directly through the roof of the third ventricle (**Fig. 31.13**). Even when an obvious CSP is absent, the midline plane of the septum pellucidum and the forniceal columns are easily identifiable and separable in children, making this a preferred route by many pediatric neurosurgeons. In older patients without CSP, this approach sometimes requires a small incision superior and parallel to the fornix (**Fig. 31.14**) to initiate the separation of the two leaves of the septum. Under these conditions, the correct midline dissection is simultaneously the most critical, as well as most challenging, aspect of this approach.

Comparison of the Two Routes

Both options rely on dissection along the fornix, and by involving both fornices, the interforniceal approach has a theoretically higher risk for postoperative cognitive problems, which has not been confirmed by convincing clinical data. But because of the position of the column of the fornix, the transchoroidal

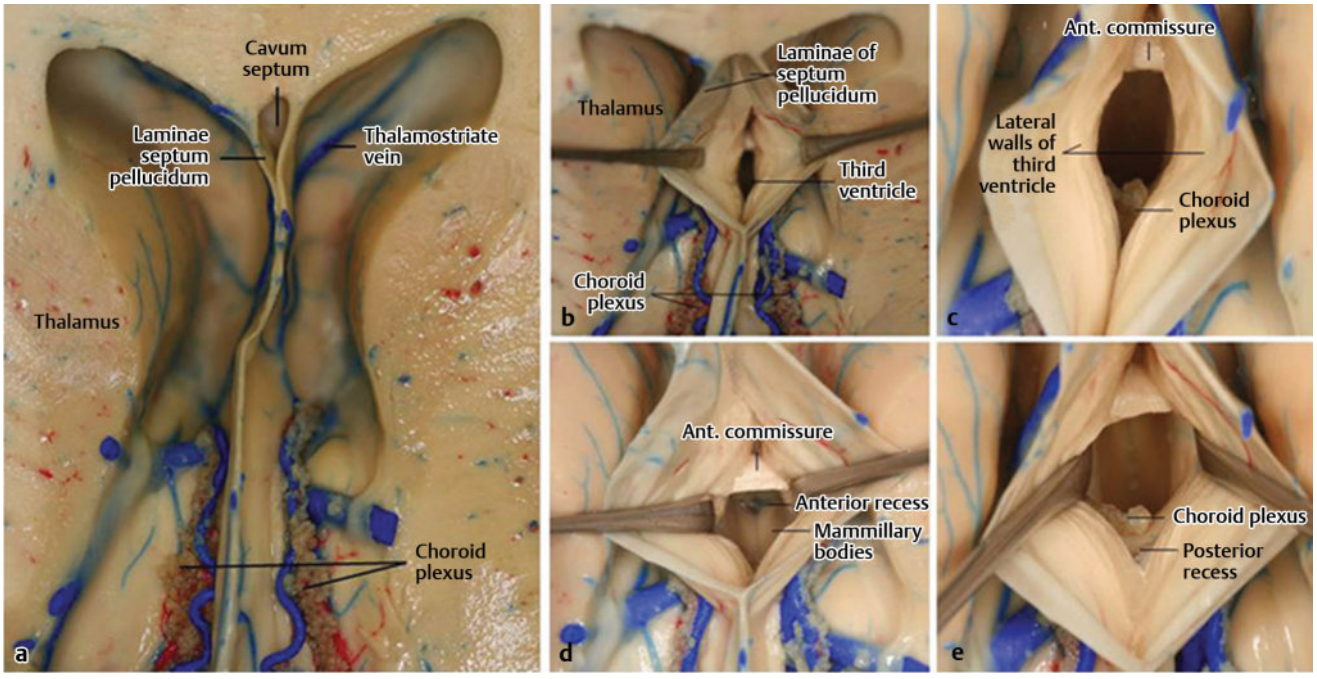


Fig. 31.13 The transcallosal anterior interforniceal approach. (a) Axial section of the adult cadaveric brain at the level of the lateral ventricle: The leaflets of septum pellucidum are going to be separated. **(b–e)** The two leaflets are gently separated, and the main structures of the third ventricle's cavity are progressively encountered. From anterior to posterior, the following structures are depicted: the anterior commissure and anterior recess, the columns of fornix, the mammillary bodies, the choroid plexus, and the posterior recess. (Reproduced from Graziano F, Ganau M, Meccio F, et al. The Transcallosal Anterior Interforniceal Approach: A Microsurgical Anatomy Study. Journal of Neurological Surgery Part B Skull Base. 2015; 76(03): 183–188.)

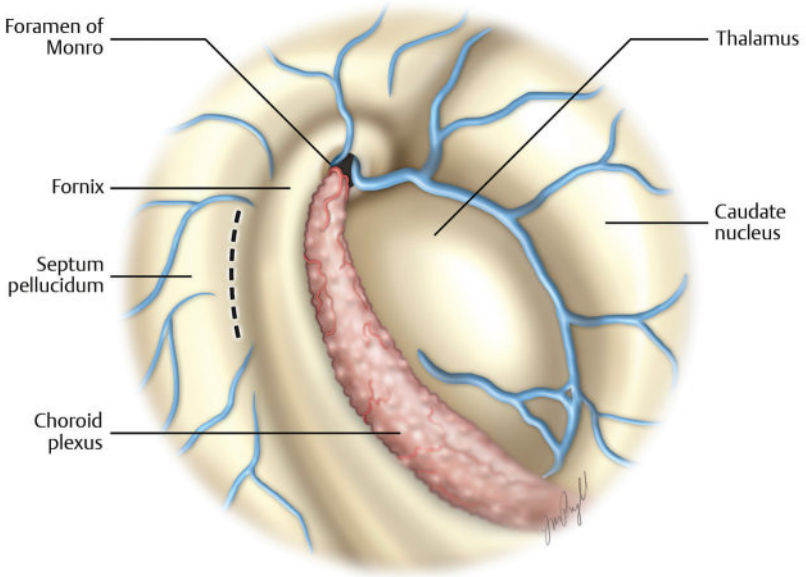


Fig. 31.14 Incision along the fornix. When a cavum septum pellucidum is absent, the interforniceal approach may require an incision along and superior to the body of the fornix in the midline. This step is demonstrated starting from the right lateral ventricle in this illustration (*dashed line*). The leaflets of the septum and the fornices can then be separated to access the third ventricle.

approach has limited exposure of the anterior region of the third ventricle, and the interforniceal approach is a true straight midline approach and offers better visualization of the entire floor of the third ventricle, with a larger work area for more surgical freedom. Its major drawback is, in the absence of a CSP, separation of the two leaves of the septum pellucidum can be technically challenging.

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32 Fourth Ventricle

David J. Daniels and Cody L. Nesvick

Keywords: tela choroidea, vermis, mutism, ependymoma, cavernous malformation, hemangioblastoma

■ Case Presentation

An 18-month-old boy presented with a 3-week history of vomiting, fussiness, and clumsiness. On initial evaluation, he was awake, alert, and interactive. His only neurologic deficit was diffuse, moderate limb ataxia. A brain magnetic resonance imaging (MRI) was obtained which demonstrated a large mass filling the fourth ventricle, causing obstructive hydrocephalus with transependymal flow (**Fig. 32.1, Fig. 32.2**). The boy was therefore admitted for treatments.

Questions

1. What is the child's most acute problem and what intervention is required right away?
2. What other studies are necessary to complete the diagnostic assessments?
3. What is the differential diagnosis of the fourth ventricular mass in this boy?

■ Diagnosis and Assessment

The MRI showed a large extra-axial lesion, measuring 4.2 × 3.4 × 4.7 cm, which exerted significant mass effect on the cerebellum, midbrain, and pons (**Fig. 32.1, Fig. 32.2**). The mass was heterogeneous on T1- and T2-weighted images and generally hypointense compared to white matter on T1. There was some patchy contrast enhancement within the mass but no restricted diffusion. The lesion extended out the foramina of Luschka bilaterally, wrapping around the medulla, lower cranial nerves, and posterior inferior cerebellar artery (PICA) (**Fig. 32.1a–c**).

Taking together the tumor location, lack of restricted diffusion, and a history of vomiting, the most likely diagnosis was an ependymoma (**Fig. 32.3**). Other tumors on the differential diagnosis are medulloblastoma (**Fig. 32.4**) and other embronyal types of tumor. A medulloblastoma would be considered more seriously had the overall bulk of the lesion been situated higher in the fourth ventricle and had restricted diffusion (**Fig. 32.4**). Other embryonal tumors, such as atypical teratoid rhabdoid tumor are also in the differential diagnosis.

The first consideration was whether to place an external ventricular drain (EVD). At our institution, the practice is to place an EVD for obstructive hydrocephalus prior to definitive surgery. There is always theoretical concern for upward

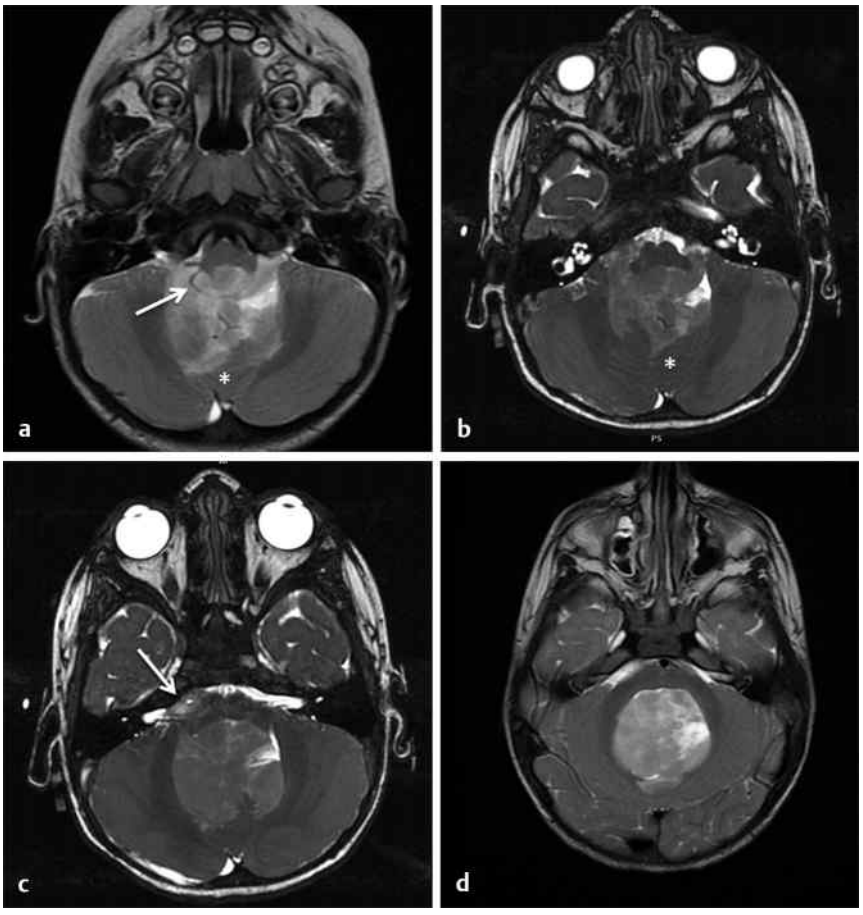


Fig. 32.1 Preoperative MRI of an 18-month-old child with vomiting and ataxia. (a) T2-weighted axial scan showed how the lesion came out of the fourth ventricle through the foramina of Luschka with complete encasement of the right posterior inferior cerebellar artery (*arrow*); (*) highlights the vermis. **(b)** Axial FIESTA imaging showed how the lesion wrapped extensively around the lower brainstem bilaterally. **(c)** Axial FIESTA imaging continued to show tumor in from the brainstem on the right (*arrow*). **(d)** Axial T2 scan showed how it filled the fourth ventricle.

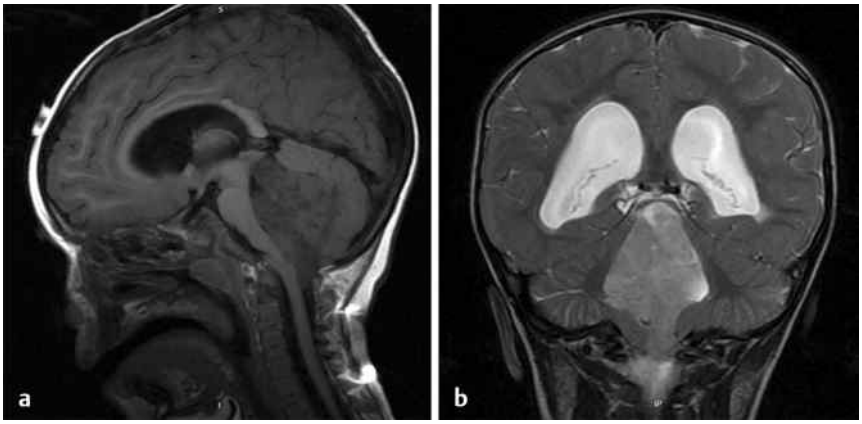


Fig. 32.2 Preoperative images. (a) Sagittal T1 image showed how the lesion extended from the C1 lamina to the cerebral aqueduct and (**b**) T2 coronal scan illustrated how the tumor filled the entire fourth ventricle.

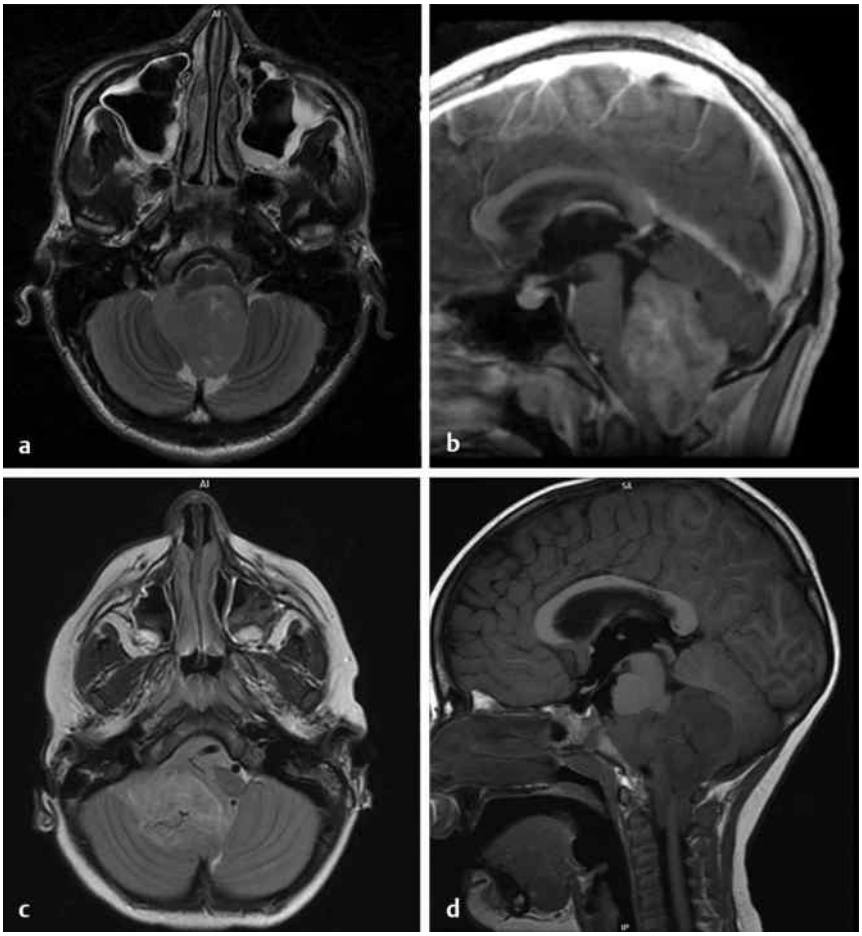


Fig. 32.3 Other ependymoma cases. (a, b) A large fourth ventricular ependymoma that exited the ventricle through the right foramen of Luschka. This boy had a 3-month vomiting history and his tumor was very adherent to the area postrema. **(c, d)** MRI showed an ependymoma with an extended component in the right cerebellopontine angle that encased all the lower cranial nerves and blood vessels. This lesion was approached with a midline incision and telovelar approach.

herniation, but with judicious cerebrospinal fluid (CSF) drainage, this is rarely clinically consequential. We generally maintain our EVDs open at a fixed pressure of at least 13 cm H_2O or 10 mm Hg prior to surgery to facilitate ventricular access, and drain additional CSF intraoperatively as needed. Additionally, we advocate obtaining full neuraxis imaging prior to surgery for the purposes of preoperative staging. Since this study requires sedation in an 18-month-old boy, it is much safer to perform the study with an EVD in place.

In the case presented here, an EVD was placed on admission, and full imaging including stealth MRI was obtained. Fortunately, the spine MRI did not reveal any "drop" metastasis. From a surgical perspective, the goal of surgery for a large fourth ventricular lesion in a young child is to obtain a gross total resection (GTR). Given their tendency to adhere to the floor of the fourth ventricle and involve cranial nerves, ependymomas can be very challenging to resect. Nevertheless, the degree of surgical resection is closely linked to prognosis in ependymoma and certain forms of medulloblastoma, so maximal safe surgical resection is necessary. So strong is the evidence for this that second-look surgery is often necessary, if tumor residual were discovered after resection of an ependymoma or medulloblastoma.

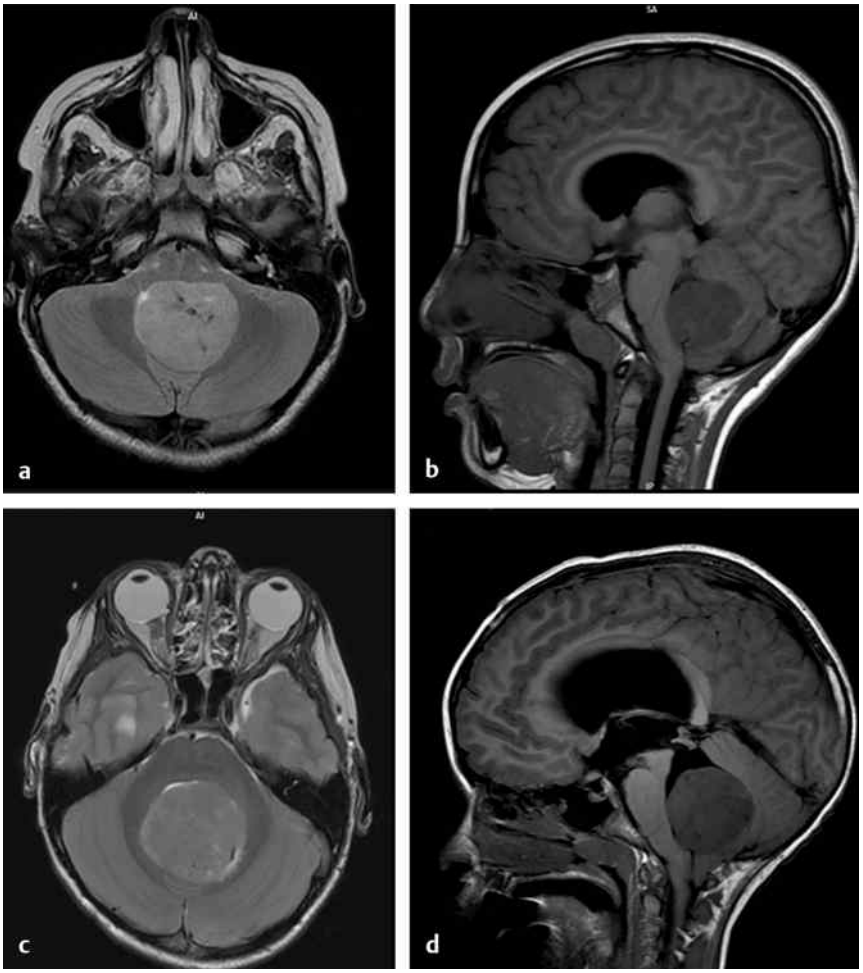


Fig. 32.4 MRI of two medulloblastomas. These images highlight the general concept that these lesions do not usually extend out the foramina of Luschka and are contained in the fourth ventricle. **(a)** Axial and **(b)** sagittal MRI in a 4-year-old boy who presented with headache, nausea, and vomiting from obstructive hydrocephalus and found to have a large fourth ventricular lesion confined to the fourth ventricle. The tumor had restricted diffusion (not shown). **(c)** Axial and **(d)** sagittal MRI in a 2-year-old boy with similar symptoms showed similar imaging. Both tumors turned out to be medulloblastomas.

■ Anatomical and Therapeutic Considerations

A thorough knowledge of fourth ventricular anatomy is necessary for surgery in this area of the brain, and the local topography must serve as a guide to safe resection. As in many cases, the bulk of this patient's tumor filled the fourth ventricle, seemingly simple to resect. However, many of these tumors would prove to be adherent to the floor of the fourth ventricle, which includes the vagal trigone, hypoglossal trigone, and facial colliculus. In our patient, the tumor wrapped around the medulla, the lower cranial nerves, and encased the right PICA (**Fig. 32.5**). The superior and inferior cerebellar peduncles, which comprise the lateral walls of the fourth ventricle, must be recognized on the MRI, as they represent the lateral limits of the tumor resection.

Since the majority of the tumor filled the fourth ventricle, a midline approach is required. However, this tumor also extended out both foramen of Luschka, wrapping around the lateral side of the brainstem (**Fig. 32.1a, b**). Had this lateral portion existed in isolation, a retrosigmoid approach would have been best suited for it. Accessing the lateral portion through a midline approach, as mandated by our large tumor, would require the elevation of the cerebellar tonsils. Finally, the right medullary segment of PICA **(Fig. 32.1a**) was clearly engulfed in the tumor and planned sacrifice must be considered.

Options for Approach

In order to remove large tumors in the fourth ventricle, wide exposure into the ventricle must be obtained. In the past, this kind of wide ventricular access was almost exclusively obtained by removing part of a cerebellar hemisphere or by splitting the cerebellar vermis. The two most common surgical approaches used today to the fourth ventricle are the transvermian approach and the telovelar approach. The more traditional transvermian approach gives the surgeon good access to the entire fourth ventricle, whereas the telovelar approach provides more optimal exposure laterally. The technical differences between the two approaches, and the anatomical structures that limit the surgical view in each approach, lead to differences in the exposure of the fourth ventricle floor, the lateral recess, and the foramen of Luschka.

The transvermian approach consists of splitting the inferior vermis via an incision that extends through the uvula, pyramid, tuber, and folium of the vermis, depending on size of the pathological area (**Fig. 32.6**). Functionally, the decussating fibers of the superior cerebellar peduncle lie deep in relation to the superior medullary velum, which is a thin lamina of white matter between the superior cerebellar peduncles. The vermian incision exposes the underlying nodule, which is necessarily incised along with the tela choroidea and inferior medullary velum to gain access to the fourth ventricle. Retraction of the two halves of the lower vermis provides approximately 1 to

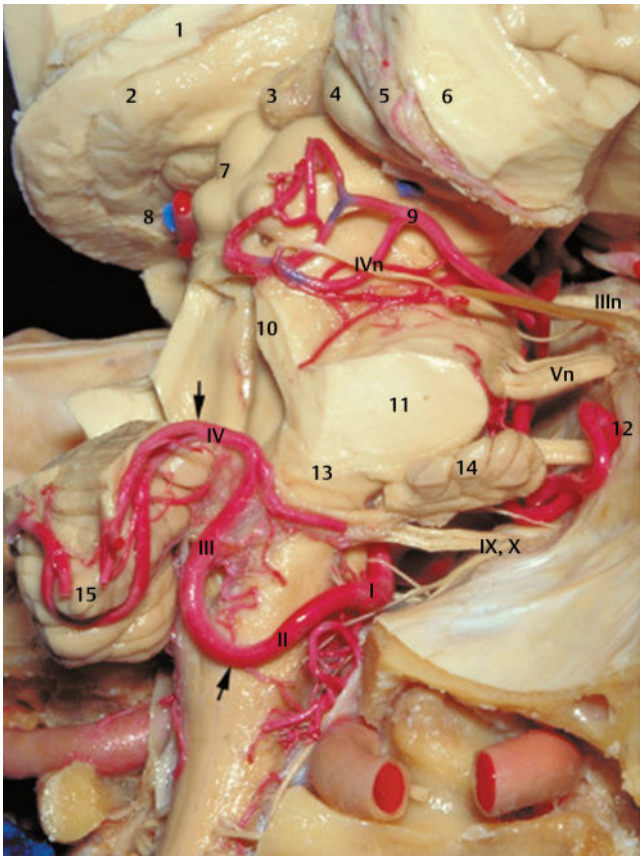


Fig. 32.5 Posterolateral view of the right posterior inferior cerebellar artery (PICA) to display its relationship to the fourth ventricle. 1, fornix; 2, pulvinar of the thalamus (cisternal); 3, pineal; 4, pulvinar of the thalamus; 5, choroidal fissure; 6, pulvinar of the thalamus (atrial); 7, quadrigeminal plate; 8, basal vein and PCA; 9, superior cerebellar artery; 10, superior cerebellar peduncle; 11, middle cerebellar peduncle; 12, internal acoustic meatus, anterior inferior cerebellar artery, facial and vestibulocochlear nerves; 13, inferior cerebellar peduncle; 14, flocculus; 15, pyramid; IIIn, oculomotor nerve; IVn, trochlear nerve; Vn, trigeminal nerve; IX, X, glossopharyngeal and vagus nerves. I, anteromedullary segment of PICA; II, lateromedullary segment of PICA; III, posteromedullary segment of PICA; IV, supratonsillar segment of PICA. Caudal and cranial loops (*arrows*). (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

2 cm of working space between the two edges of the inferior vermis. Anatomic studies show this provides excellent working room and exposure to pathologies located in the midline fourth ventricle all the way up to the aqueduct. The major risks of this approach are damage to the vermis and decussating fibers of the superior cerebellar peduncle if the incision is carried too far superiorly, which result in cerebellar mutism and paresis, respectively. Cerebellar mutism may not appear until days after surgery, and may take weeks or months to resolve.

By contrast, using the telovelar approach, the tela choroidea and inferior medullary velum, which form the lower half of the roof of the fourth ventricle, are opened on either side of the vermis to provide exposure into the fourth ventricle (**Fig. 32.7**). This approach exposes the full extent of the floor

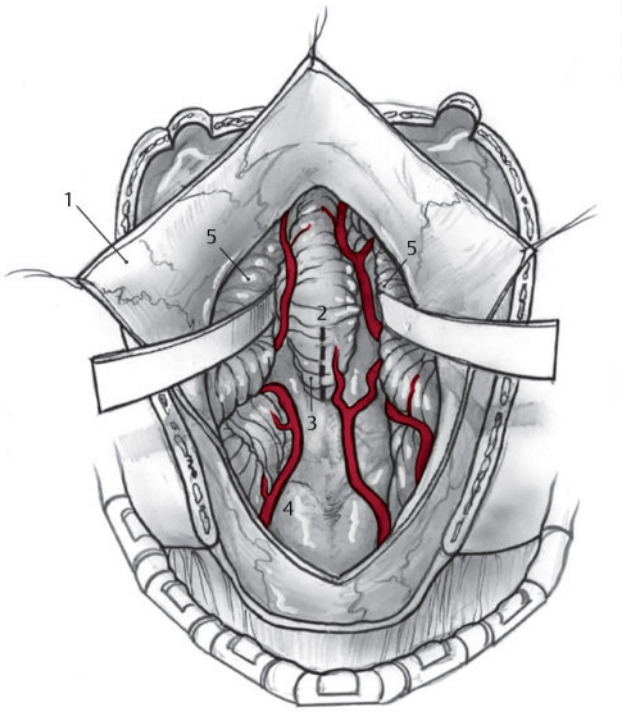


Fig. 32.6 Illustration of the transvermian approach. The cerebellar vermis is incised to gain access to the fourth ventricle in this approach. 1, dura; 2, vermis; 3, uvula; 4, obex; 5, cerebellar hemispheres. (Reproduced from Nader R et al, eds. Neurosurgery Tricks of the Trade. Thieme; 2014.)

of the fourth ventricle in a manner similar to the transvermian approach (**Fig. 32.7b**). In addition, structures along the lateral recess of the fourth ventricle, including the foramen of Luschka, can be accessed without removal of cerebellum or adjustment of retractors. This approach is excellent for lesions in the lateral recess and ones that extend through the foramen of Luschka.

The transvermian and telovelar approaches differ in several key aspects. These include the direction and route of the approach, the structures retracted and sacrificed, the direction of the retraction, the limitation of the exposure, and the areas exposed. Selection between these approaches should be tailored to the location and direction of the growth of the fourth ventricular or brainstem lesion, for which surgery is being designed. Lesions located in the vermis and extending into the fourth ventricle are best accessed via the transvermian approach, whereas tumors located on the ependymal surfaces and in the floor of the fourth ventricle often do not invade the cerebellum and can be accessed using the telovelar approach.

Approach of Choice

The telovelar approach was selected for our patient because his lesion extended laterally on both sides and wrapped around the medulla bilaterally. Freeing the arachnoid along the tonsillomedullary interface bilaterally would expose the lateral recesses of the fourth ventricle from a midline approach. The lower rate of cerebellar mutism and other cerebellar deficits from this approach make this more attractive.

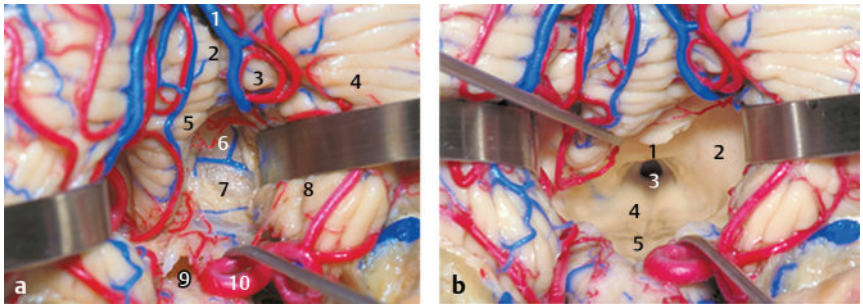


Fig. 32.7 Anatomical dissection displaying the telovelar approach to the fourth ventricle. (a) The right tonsil has been "freed" by dissecting the spaces around it (medial, anterior, and superior spaces) and retracted laterally. 1, inferior vermian vein; 2, pyramid; 3, copula pyramidis; 4, biventral lobule; 5, uvula; 6, inferior medullary velum; 7, tela choroidea; 8, tonsil; 9, foramen of Magendie; 10, posterior inferior cerebellar artery. **(b)** View of the fourth ventricle offered by the telovelar approach. The exposure is clear to the level of the aqueduct. The right tonsil has been removed for illustrative clarity; the uvula has been retracted superiorly and the left tonsil retracted laterally. 1, superior medullary velum; 2, superior cerebellar peduncle; 3, aqueduct; 4, facial colliculus; 5, stria medullaris of the fourth ventricle. (Reproduced from Sekhar L, Fessler R, eds. Atlas of Neurosurgical Techniques. Brain. 2nd ed. Thieme; 2016.)

Questions

1. What is the superior limit of the telovelar approach?
2. Classically described, where is the cut that opens the tela choroidea and inferior medullary velum?
3. How far laterally does one proceed with this cut?

■ Description of the Technique

As with any craniotomy, positioning is critical to facilitate proper exposure, maximize surgical freedom, and minimize neurological complications. For fourth ventricular lesions, neck flexion is necessary to obtain the necessary view of the cerebral aqueduct, but it is important not to overflex the neck and occlude jugular venous outflow, which could exacerbate cerebral edema and bleeding during surgery. Because of the patient's young age and likely long-standing hydrocephalus, we opted not to use Mayfield fixation. The upper forehead and vertex were rested on a foam-padded horseshoe head holder which was positioned below the torso to accommodate neck flexion. In this manner, the chin was tucked significantly allowing the surgeon the best access to the superior part of the fourth ventricle (see **The Three Approach Elements**).

The use of neuromonitoring is debatable, as we needed to remove this tumor in its entirety regardless of signal changes. However, we routinely use neuromonitoring of cranial nerves V, VII, X, and XI, in addition to motor evoked potentials (MEPs) and somatosensory evoked potentials (SSEPs), if we are worried about vascular events around the brainstem. Hypoglossal nerve monitoring can result in significant tongue swelling in children, which can delay extubation. It is therefore seldom used in our practice. Monitoring the vagal nerve requires electrodes on the endotracheal tube and an anesthesia team familiar with this technique.

The Three Approach Elements

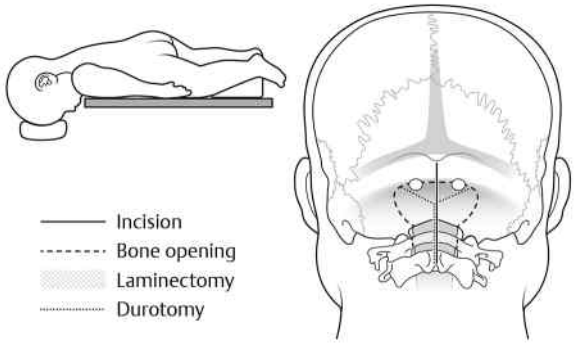
Corridor: subtonsillar
Craniotomy: suboccipital
Modifier: incision at telovelar junction

Although not critical for this tumor, neuronavigation was used nonetheless. A midline incision was started above the level of the inion and carried down to the C2 spinous process. The avascular plane in the midline was developed to separate the muscles. Superiorly, the bone over the inion was exposed, while inferiorly, the ring of C1 was dissected clear a little wider than the width of the spinal canal (see **Operative Setup**).

The suboccipital craniotomy was then made, starting with two burr holes on either side of the bony keel, using navigation guidance to make sure the holes are at least 3 cm above foramen magnum. The dura was stripped, and a Kerrison ronguer was used to connect these two holes at the keel. The atlantooccipital membrane was stripped from foramen magnum using an angled curette. Laterally, notches were made with rongeurs on either side of the foramen magnum as "landing spots" for the craniotome footplate. These notches were spaced wide enough

Operative Setup

Position: prone
Incision: midline, inion to C2
Bone Opening: midline suboccipital
Durotomy: Y-shaped



apart to ensure full exposure of the tumor extending around the lower brainstem. The craniotomy was made by using the footplate laterally from each burr hole, going as lateral as necessary, before turning inferiorly toward the foramen magnum exiting the notches that was previously created. This created a one-piece craniotomy that can be easily reattached at the end of the case.

Although a C1 laminectomy is not routinely used with the telovelar approach, it was necessary here because our tumor extended inferiorly to the C1 arch. For even wider exposure at the foramen magnum, rongeurs were used to extend the bony opening to the occipital condyle bilaterally.

For a midline exposure to the fourth ventricle a standard Y-shaped dural opening is sufficient. We started over the midline spinal dura below the level of C1 and inferior to both PICAs where they could be "pinned" back towards the dura at the cervicomedullary junction. CSF was drained from the cisterna magna. The dural opening was then carried superiorly to foramen magnum and then the two upper limbs of the "Y" were made by angling laterally in each direction. It was important to go slowly with the dural opening as a circular sinus, occipital sinus, and venous lakes can be extensive in this region, especially in children.

The tumor was found extending out of the fourth ventricle, and several pieces were removed for histopathological analysis. The diagnosis from this was ependymoma. The "telovelar" approach was so named because it traverses the tela choroidea and inferior medullary velum, which together form the inferior half of the roof of the fourth ventricle (**Fig. 32.7**). First, the arachnoid around the cerebellar tonsils were sharply incised with an arachnoid knife. To facilitate lateral retraction of the tonsils, the arachnoid between the tonsils and vermis was incised, and the plane between them was carefully opened one side at a time. With deeper dissection, the anterior surface of the tonsils was visualized, exposing choroid plexus, choroidal vessels, and a thin membrane, which is the tela choroidea. The telovelar junction, the line of attachment of the inferior medullary velum to the tela choroidea, extends from the vermis to each lateral recess. Using bipolar cautery, the vessels and choroid plexus were coagulated, and the membrane was then opened with micro-scissors. Performing this technique bilaterally increases the opening to the fourth ventricle substantially (**Fig. 32.8**). With the exception of an initial biopsy, we generally respect tumor margins and do not debulk these tumors until exposure is completed, as this may lead to untoward bleeding.

The bulky portion of the lesion that filled the entire fourth ventricle was removed without difficulty, as the tumor separated nicely along the floor of the fourth ventricle and the superior medullary velum. Ependymomas usually respond favorably to suction removal and this patient's tumor was no exception. The cerebellar tonsils were mobilized superiorly and laterally to expose the lateral side of the medulla, and the tumor's lateral extensions were safely resected. However, the right medullary segment of

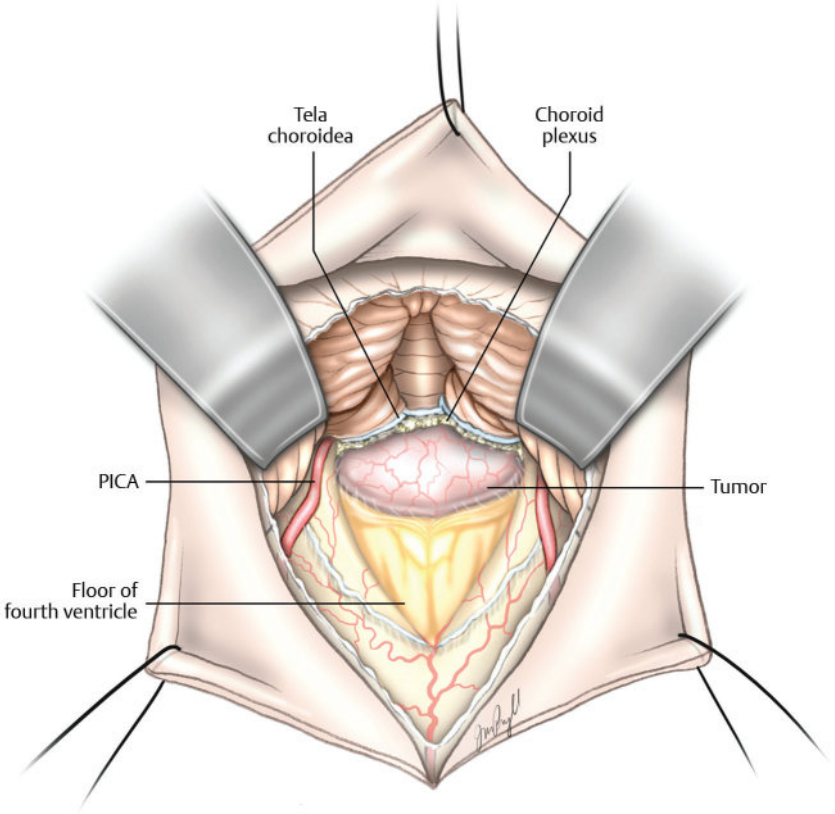


Fig. 32.8 Tumor exposure through the telovelar approach. The telovelar junction is incised on both sides and the cerebellar tonsils are mobilized superiorly. With this, exposure to the tumor in the fourth ventricle is gained. PICA, posterior inferior cerebellar artery.

the PICA was encased by the tumor, and as we disentangled the vessel, tumor cells remained attached to the arachnoid along the course of the PICA. As such, the vessel was sacrificed to obtain a total resection. At the end of the operation, meticulous inspection was made around the brainstem and cranial nerves to make sure no tumor was left behind.

Surgical Pearls

1. The telovelar approach involves opening the telovelar junction on the inferior half of the roof of the fourth ventricle. A common misconception regarding this approach is that approaching lesions extending into the upper half of the fourth ventricle is difficult because of a sharp working angle and limited exposure. In fact, lesions that extend all the way to the cerebral aqueduct are reachable through this approach. As it involves less destruction of cerebellar tissue, the telovelar approach is associated with better surgical results and fewer complications than the transvermian alternative.
2. To take full advantage of the strengths of this approach, the incision at the telovelar junction must be taken widely towards the lateral recesses on each side. This naturally affords more lateral exposure but, as it frees up the tonsils more extensively from their inferior "anchor," it also increases the superior exposure of the approach as well.

■ Aftercare

Overall, the patient tolerated surgery well. His neurologic exam was notable for a trace right-sided hemiparesis and ataxia, but he was otherwise intact. His incisional pain was well-tolerated on a standard postoperative pain regimen. His EVD was quickly weaned following the case and removed. Postoperative MRI obtained the day after surgery confirmed a GTR (**Fig. 32.9**). There was also restricted diffusion along the medial–inferior aspect of the right cerebellar hemisphere consistent with a small PICA infarct. This was not unexpected as the artery had to be sacrificed for a potentially curative resection. This idea was thrown into doubt, however, as the definitive pathological analysis showed a grade III anaplastic ependymoma.

He did very well from his craniotomy and made a very good recovery. He went on to have focal proton radiation to his posterior fossa.

■ Possible Complications and Associated Management

The overall risks for a telovelar approach for fourth ventricular tumor resection are quite high as a GTR is often necessary to prolong survival. This is especially true in ependymomas, for

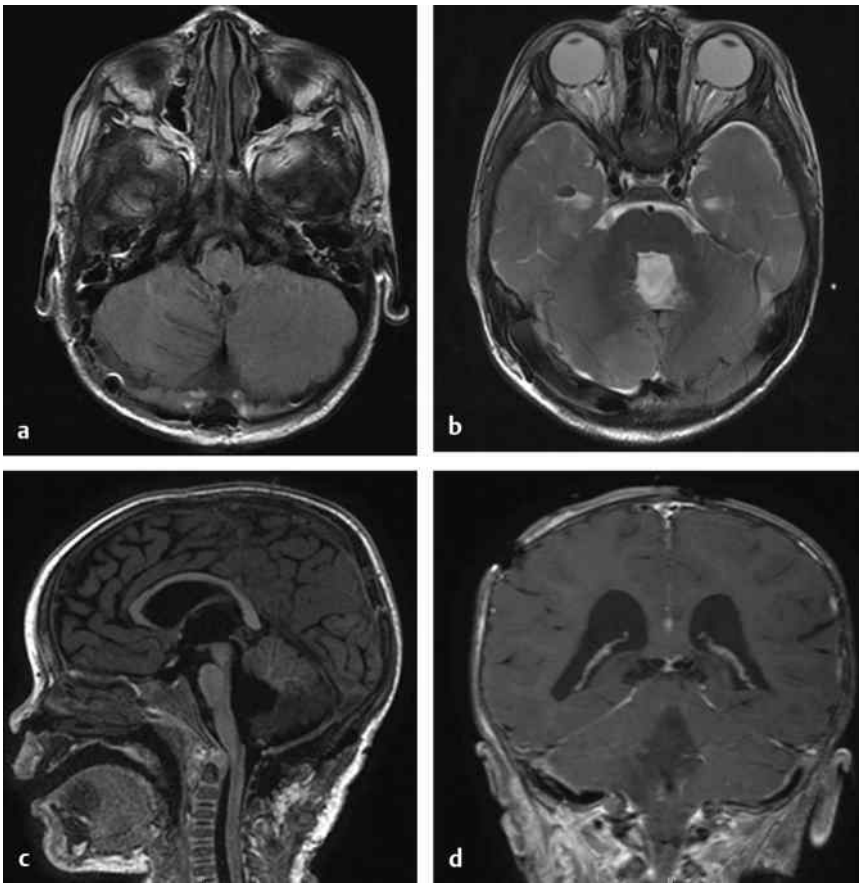


Fig. 32.9 Postoperative MRI. A gross total resection of the fourth ventricular ependymoma was achieved. **(a)** Axial T1-imaging showed tumor around the lower brainstem has been removed. **(b)** Axial T2-imaging confirmed good resection in the fourth ventricle and developing T2 signal in the medial right cerebellum with corresponding restricted diffusion consistent with right posterior inferior cerebellar artery infarct. **(c)** Sagittal T1 imaging and **(d)** coronal T1 with gadolinium revealed excellent tumor resection from the fourth ventricle.

which there are no reliable alternatives to surgery. Due to the neural structures along the floor of the fourth ventricle and the neighboring vasculature, the overall risk for adverse outcomes following surgery is more than 30%, which includes bilateral sixth and seventh nerves palsies, nystagmus, hearing loss, worsening ataxia, loss of airway protection, and swallowing difficulties (potentially requiring tracheostomy and percutaneous gastric tube placement). One of the most feared complications of posterior fossa surgery is cerebellar mutism (also known as posterior fossa syndrome), a complex clinical entity that is characterized by lack of speech, behavioral issues, and ataxia that is not associated with long tract signs, cranial nerve palsies, and/or alteration of consciousness. Although the exact anatomical substrate for this syndrome remains unknown, the majority of complications occur after removal of midline tumors involving the vermis or in tumors that infiltrate into the cerebellar peduncles. In addition to the risk for neurologic injury, surgical site infection, CSF leak and the potential for alterations in CSF dynamics requiring a ventriculoperitoneal shunt should be discussed preoperatively with patients and their family.

Perspective

Francesco Tomasello, Filippo Flavio Angileri, Alfredo Conti, Salvatore Cardali, and Antonino Germanò

■ Introduction

Lesions located in the fourth ventricle cavity and involving its floor represent one of the most difficult neurosurgical problems. The narrow surgical corridor and proximity of vital neurovascular structures are the main limitations of operating in this area. Between the two main surgical approaches, transvermian and telovelar, the former may be more commonly used, but it requires sacrifice of functional neural tissue and carries a risk of postoperative cerebellar syndrome. As such, after the monumental anatomical studies by Rhoton, Mussi, and Matsushima, the telovelar approach, which exposes the fourth ventricle widely without destroying healthy brain tissue, has gained progressive acceptance as the favored approach to manage fourth ventricular tumors.

■ Case Presentation 1

A 42-year-old woman presented to our hospital because of two episodes of diplopia associated with headache in last 2 years. In both cases, the symptoms spontaneously disappeared after a few days. On admission, her examination showed mild right sixth nerve palsy. MRI scan revealed a small cavernoma in the fourth ventricle floor at the level of the middle cerebellar peduncle and the right lateral recess (**Fig. 32.10**). Four-vessel angiogram did not reveal any aneurysm or angiographically visible vascular malformation (**Fig. 32.11**).

Anatomical and Therapeutic Considerations

The inferior portion of the cerebellar vermis is deeply embedded between the two hemispheres. From top to bottom, the vermian surface consists of the folium, tuber, pyramid, and uvula. The uvula lies between the cerebellar tonsils. By elevating the cerebellar tonsils, the uvula, together with the tela choroidea and the inferior medullary velum, forming the lower half of the roof of the fourth ventricle, can be exposed. The tela choroidea is attached to the medulla oblongata inferolaterally via the taenia, conjoined in a V-shaped pattern. At this level,

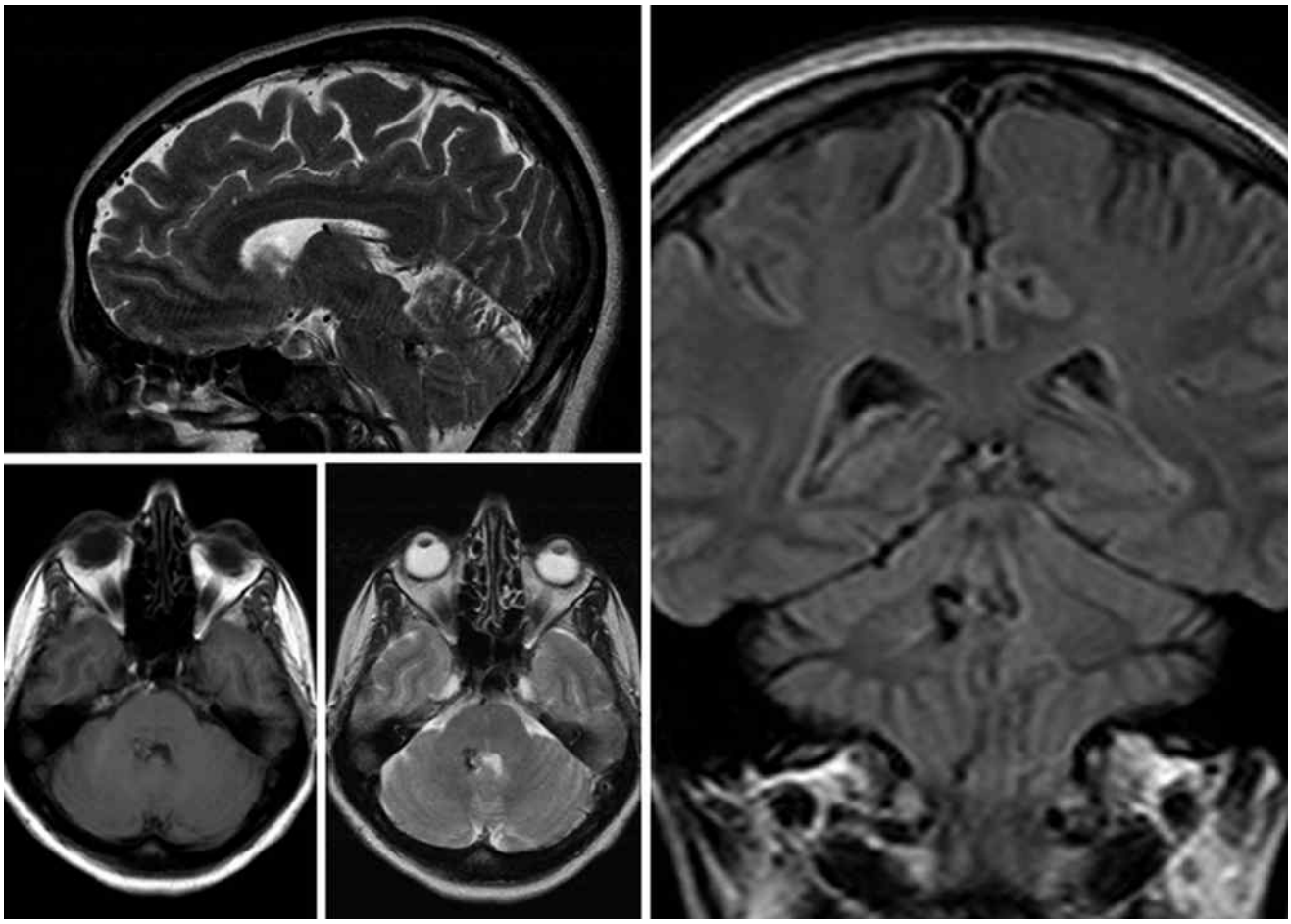


Fig. 32.10 Preoperative MRI of Case 1. These images revealed a small cavernoma in the fourth ventricle floor at the level of the middle cerebellar peduncle and the right lateral recess.

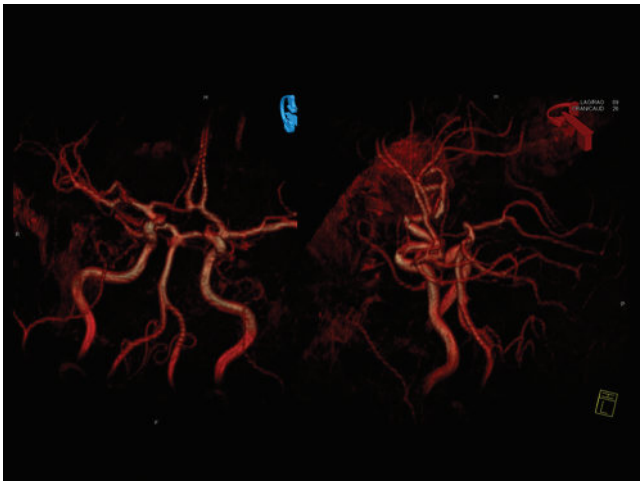


Fig. 32.11 Angiography of Case 1. Vascular imaging did not reveal clear arterial feeders or venous outflow of the brainstem malformation, consistent with the presumptive diagnosis of a cavernous malformation.

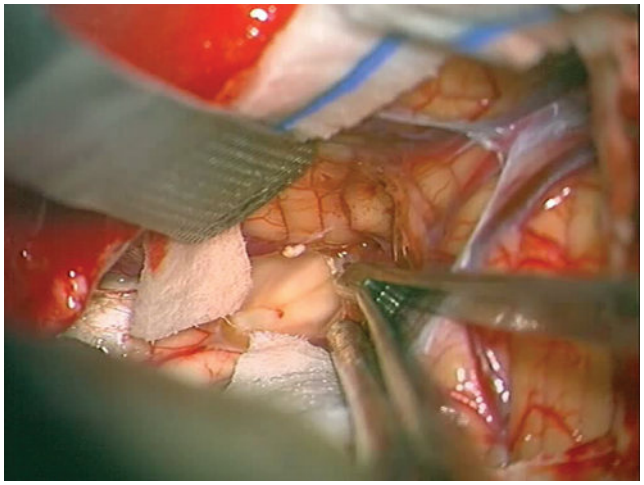


Fig. 32.12 Dissection of the tela choroidea. The inferior edges of the tonsils were exposed and the dissection extended between the medial side of the tonsil and the adjacent edge of the uvula.

the fourth ventricle communicates with the cisterna magna through the foramen Magendie. Rostrally, the taenia continues along the ventricular entrance into the lateral recess. The inferior medullary velum is a thin bilateral layer of neural tissue that extends from the nodule medially to the dorsal margin of each lateral recess, forming the peduncle of each flocculus laterally. Caudally, the inferior medullary velum attaches to the tela choroidea. In the rostral extension of the cerebellomedullary fissure, the PICA courses around the superior pole of the tonsil.

Surgical removal of cavernous malformation may be a significant challenge. The goal of surgery should always be balanced against the natural history of untreated lesions, and for asymptomatic brainstem cavernomas, the risks may outweigh the benefits. However, for symptomatic lesions, surgical treatment may be justified. When considering surgery for a pontine cavernoma with clear signs for hemorrhage, the access is a crucial aspect of the surgical strategy. In this case, the hemorrhage around the lesion reached the fourth ventricle cavity through its floor, and this provided a relatively safe entry point into the pons. The surgical strategy should include a wide and safe exposure of the entire floor to identify the exact location of this entry point.

The telovelar approach represents, in our opinion, the best strategy to reach the target to resect the lesion safely. The floor of the fourth ventricle can be visualized as soon as the dura is opened, when both tonsils are retracted. It can be widely exposed as the tela incision progresses laterally. Moreover, the telovelar corridor would provide a tangential view on the floor. Although a transvermian approach might provide sufficient exposure to the fourth ventricular cavity, the trajectory through the inferior vermis would place the floor of the ventricle behind the lesion, making it harder to protect until later on during the operation.

Description of the Technique

The patient was placed in the prone position and the head fixed in a Mayfield head holder. The neck was flexed without compromising the venous outflow. Neurophysiologic monitoring was established, including MEPs, SSEPs, and BAEPs (brainstem auditory evoked potentials). A midline occipitocervical skin incision extending from the inion to the level of the spinous process of C3 was made, exposing the occipital bone and posterior arch of C1. A standard midline suboccipital craniectomy was performed exposing the cerebellar tonsils and inferior vermis. The posterior arch of C1 was not removed and a Y-shaped dural opening was made. The arachnoid was incised and tented to the dura by means of titanium micro-clips.

The inferior edges of the tonsils were exposed and the dissection extended between the medial side of the tonsil and the adjacent edge of the uvula (**Fig. 32.12**). The surface of the tonsil that faced the uvula was retracted superolaterally toward the side of the approach, and the uvula was retracted to the opposite side to expose the tela choroidea and the inferior medullary velum. As the tela was incised, the fourth ventricle floor was progressively exposed. The tela choroidea, which forms the caudal part of the lower half of the roof of the fourth ventricle, was bilaterally incised in a caudal to rostral direction towards the inferior medullary velum. Elevation of the tonsil, division of the tela choroidea, and opening of the inferior medullary velum allowed excellent exposure of the floor and direct access to the right lateral recess.

A clear yellowish discolored area at the level of the lateral recess was identified as the entry point to remove the cavernoma (**Fig. 32.13**). The lesion was identified immediately under the hemosiderin layer and sharply dissected. Further removal of hemosiderin was not performed. A venous malformation resembling a caput medusae was also identified and left untouched (**Fig. 32.14**). The cavernoma was resected completely (**Video 32.1**).

Aftercare

The patient had an uneventful postoperative course. The sixth cranial nerve palsy gradually improved and finally resolved at 6-month follow-up. The postoperative MRI demonstrated the complete removal of the cavernous malformation while the associated venous angioma and a small hemosiderin ring could still be seen (**Fig. 32.15**).

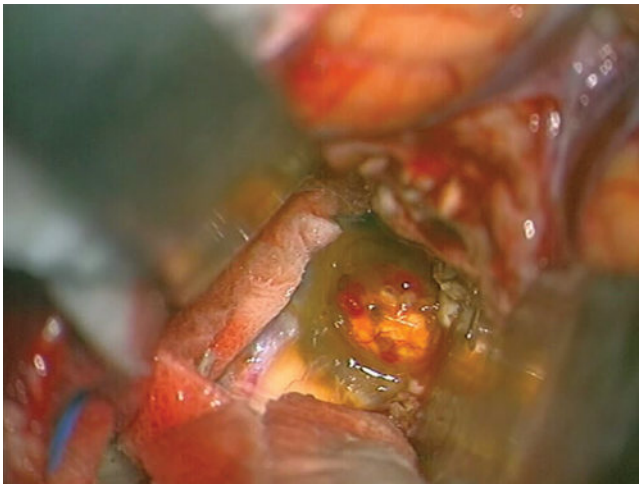


Fig. 32.13 Intraoperative image. A clear yellowish discolored area was identified on the floor of the fourth ventricle. This was the point where the lesion reached the surface of the brainstem.

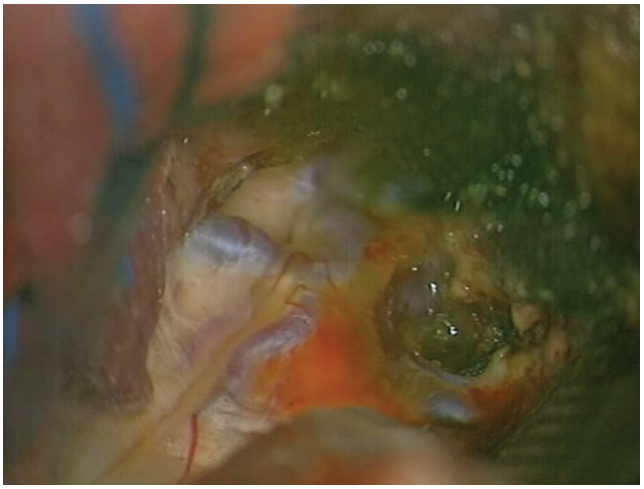


Fig. 32.14 Venous angioma. The associated venous angioma was intentionally left behind after the resection.

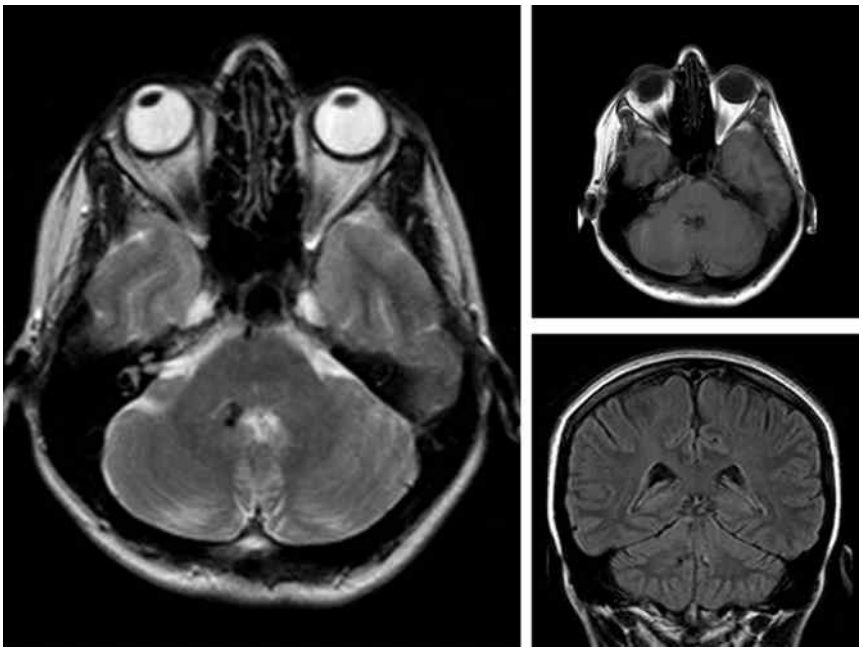


Fig. 32.15 Postoperative MRI of Case 1. A complete removal of the cavernous malformation was achieved. The venous angioma and a small hemosiderin ring was left by intention.

■ Case Presentation 2

A 35-year-old woman presented with a 15-day history of headaches, vomiting, and gait imbalance. On initial neurological evaluation, she was conscious, alert and oriented. Cranial nerve examination was normal. There were no focal motor or sensory deficits. A mild gait ataxia was evident. MRI showed a mass lesion that was hypointense on T1-weighted images and hyperintense on T2-weighted images, occupying the caudal part of the fourth ventricle and the dorsal medulla, associated with a cystic component. The caudal medulla was displaced ventrally by the cystic component. The solid portion of the lesion enhanced homogeneously after gadolinium administration. Feeding vessels to the lesion were clearly evident, and taken together, the radiologic characteristics were diagnostic for a hemangioblastoma (**Fig. 32.16**).

Anatomical and Therapeutic Considerations

Hemangioblastomas may occur either sporadically (about two-thirds of cases) or as part of von Hippel–Lindau disease (VHL about one-third). In this case, VHL disease was diagnosed after the occurrence of the cerebral hemangioblastoma.

The cystic component of the tumor was located at the level of the obex, and the solid component was superior to it, in the lower half of the fourth ventricle. A telovelar approach was our approach of choice to resect the tumor. It should be highlighted that the cyst wall of the hemangioblastoma is not neoplastic and should not be resected. Studies on the natural history of hemangioblastoma suggest that during tumor growth, cyst development occurs as peritumoral edema meets an anatomical barrier. For this reason, the surgical

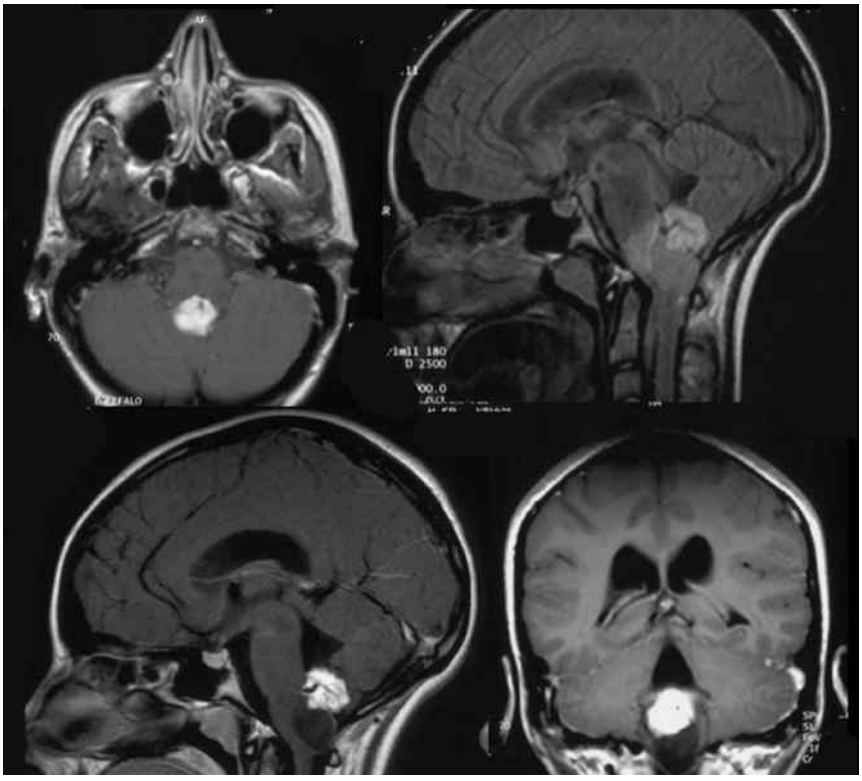


Fig. 32.16 Preoperative MRI of Case 2. This mass lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images. It occupied the caudal part of the fourth ventricle and the dorsal medulla, associated with a cystic component. The caudal medulla was displaced ventrally by the cystic component. The solid portion of the lesion enhanced homogenously after gadolinium administration.

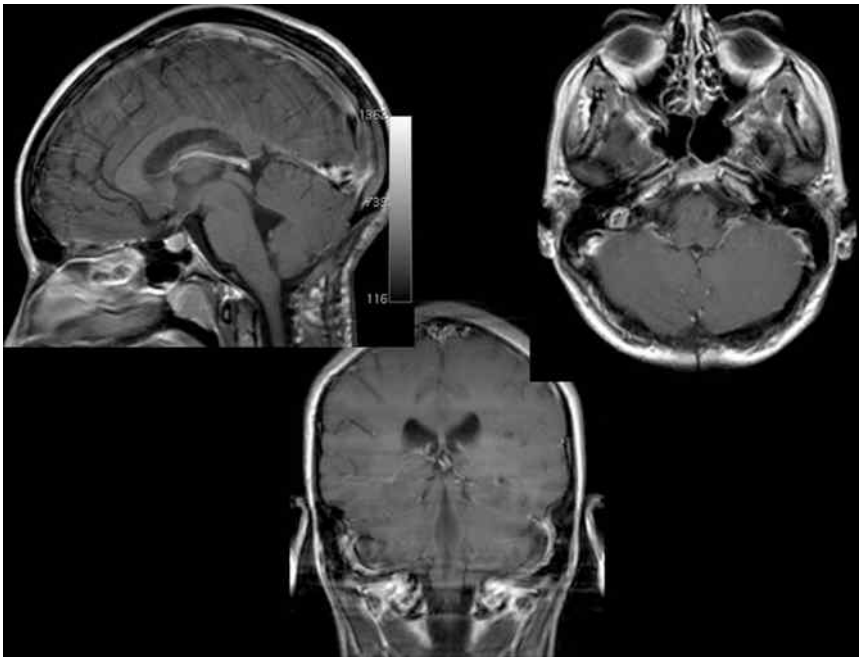


Fig. 32.17 Postoperative MRI of Case 2. These showed a complete resection of the patient's hemangioblastoma.

strategy should be aimed at the resection of the solid portion of the tumor with fenestration of the cyst.

Description of the Technique

For this case, patient positioning and skin incision were the same as in the first case. An EVD was inserted and neurophysiological functions were monitored. The posterior arch of C1 was removed to allow a better exposure of the cyst. After dural opening, the cyst expanding the medulla oblongata was exposed. The cystic wall was incised and the medulla decompressed. After arachnoidal dissection, both tonsils were laterally mobilized, exposing the solid and vascular tumor. With a hypervascular tumor, an internal debulking was not advisable and thus, it was dissected circumferentially. The small feeding arteries originating from the PICA were identified and divided, allowing step

by step mobilization of the nodule. The PICA was carefully protected. The tumor was very adherent to the floor of the fourth ventricle, and a dissection plane was developed meticulously. Finally the tumor was removed en bloc (**Video 32.2**). The pathological analysis confirmed a hemangioblastoma.

Aftercare

The postoperative course was uneventful. The ventricular drain was kept in place for 48 hours and then removed. Postoperative imaging revealed the complete removal of the tumor, and no further evidence of the cyst. Edema was significantly reduced (**Fig. 32.17**). The gait disturbances resolved at three months follow-up and the patient was enrolled in a nationwide program to manage VHL disease.

Commentary

Walter C. Jean

In lieu of the usual commentary section prevalent throughout the book, the *Perspective* authors in this chapter posed a series of questions. Although these are no doubt aimed at the reader, it does provide a unique opportunity for the editor to interject some comments.

Do you agree with the decision to excise the pontine cavernoma?

With clinical evidence of repeated hemorrhage, the decision to resect the cavernoma is sound, *as long as* the approach does not destroy healthy tissue. In this case, the lesion and related hemorrhage reached the surface of the brainstem, making the choice of entry point into the lesion quite straightforward. The approach to the brainstem itself must follow natural dissection planes to avoid neurological damage, and the telovelar approach was the best choice.

Do you agree that preoperative embolization of posterior fossa hemangioblastomas may be harmful?

Embolization of posterior fossa hemangioblastomas has been associated with patient death following the procedure. In some cases, this was due to hemorrhage of the tumor, presumably from venous obstruction, causing acute brainstem compression. Once again, surgical exposure is critical. If wide exposure can be achieved without destruction of surrounding tissue, it is safer to rid of feeding vessels with microsurgical technique rather than with embolization. In this *Perspective*, the telovelar approach provided such an exposure.

Do you believe different surgical corridors could allow for a better management of the lesions?

For tumors that grow from the superior vermis extending secondarily into the rostral third of the fourth ventricle, the transvermian approach may be preferable, but beyond those select tumors, the telovelar approach has become the "standard bearer" for accessing the fourth ventricle. The tenets of modern skull base surgery endorse the use of natural anatomic cleavage planes over cutting through healthy brain tissue to assess surgical targets, especially when the two alternatives offer equivalent exposure and maneuverability of surgical tools. And indeed, the parity of the telovelar and transvermian approaches is not only backed up by anecdotal and experiential data, but also by carefully recorded measurements. Beyond that, the telovelar approach offers the additional advantage of earlier identification and dissection of the interface between the lesion and the floor of the fourth ventricle, which in turn, leads to better protection of the brainstem, so critical for the avoidance of complications.

However, even as a "nondestructive" approach through a natural anatomical plane, the telovelar technique still has its limitations and weaknesses.

■ Case Presentation

A 34-year-old woman presented with a 2-month history of intermittent numbness over the right side of her face. When she started experiencing blurry vision in her right eye, it prompted an MRI scan (**Fig. 32.18a**). This revealed a mass lesion filling the fourth ventricle and hydrocephalus.

An EVD was placed right away to control her hydrocephalus. Given the patient's young age, otherwise good health, and the possibility of a benign tumor, the surgical goal was set for complete removal of her fourth ventricular tumor. Therefore, a telovelar approach was performed to resect the tumor. Bilateral telovelar junctions were incised widely, and the entire fourth ventricle was exposed.

The tumor had a white color and was relatively avascular. Pathological analysis yielded the diagnosis of an ependymoma. The tumor was found to be adherent to the floor of the fourth ventricle at several points, and these areas were carefully dissected in order not to harm the brainstem directly underneath. The resection was taken all the way up the aqueduct, which was free from any blockage at the end of the procedure. With a satisfactory hemostasis and no changes detected on intraoperative neuromonitoring data, the operation was halted. Closure was performed by routine.

After the operation, the patient was neurologically unchanged, but her MRI scan showed a large residual which was stuck to the roof of the ventricle (**Fig. 32.18b, c**). This portion of the tumor was completely hidden during the telovelar approach, and had been left behind. This situation presented a clinical dilemma. The patient had just gone through a big operation, well-designed and fairly executed, which had led to no complications up to this point. However, the surgical goal had not been achieved. Given the likely diagnosis of a grade I tumor, the two existing options were "watchful waiting," versus a second operation to reach the surgical goal originally set. The patient opted for the latter.

Assuming the anatomy did not change during the interim 4 days, we decided to shift the surgical approach to a transvermian route, as we predicted that we would have the same problem seeing the rostrodorsal tumor component if we returned through the telovelar approach. During the second operation, the lower half of the vermis was coagulated and

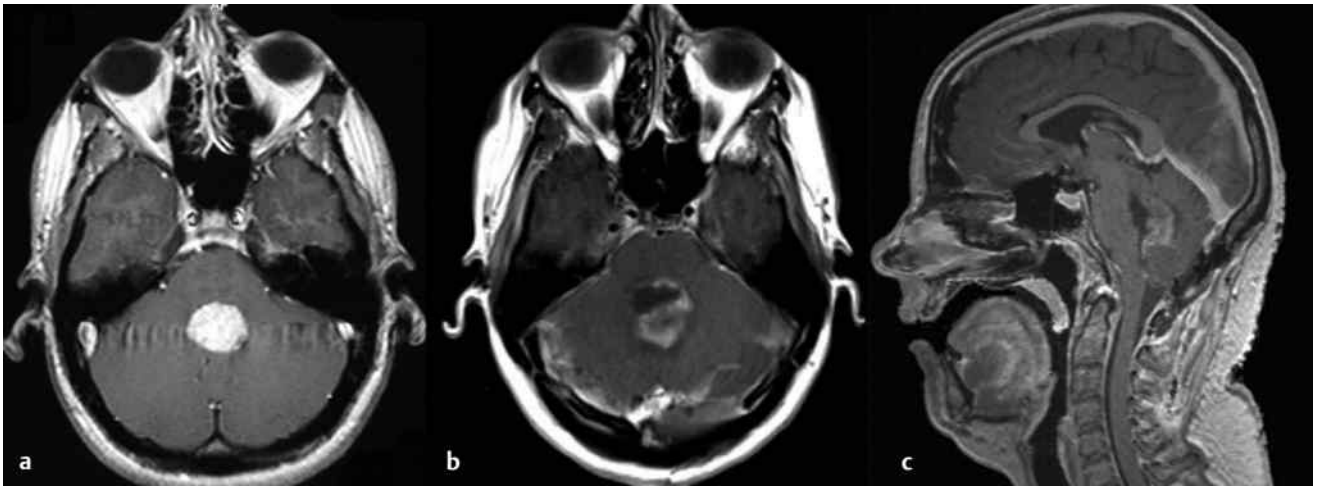


Fig. 32.18 MRI before and after telovelar surgery. (a) Axial image with gadolinium showing tumor of the fourth ventricle before surgery. **(b)** Axial image after telovelar surgery showing residual tumor attached to the roof of the fourth ventricle. The tumor at the floor had been removed. **(c)** Sagittal image after telovelar surgery showing the same residual as in **(b)**.

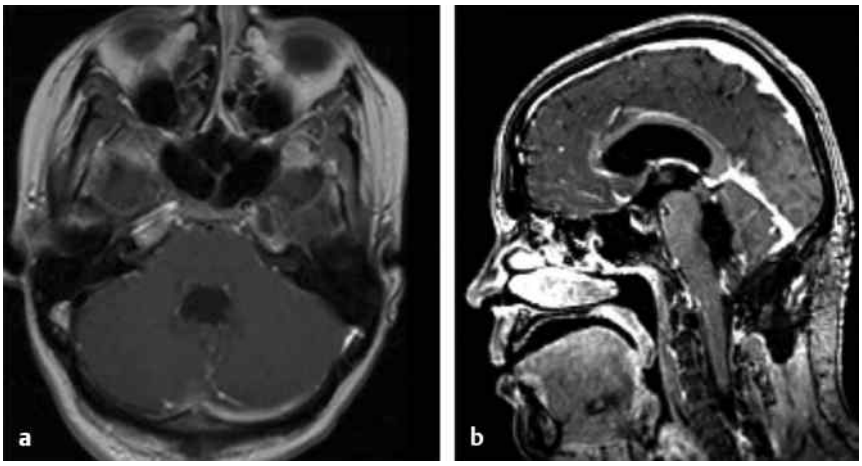


Fig. 32.19 MRIs after transvermian surgery. (a) Axial and **(b)** sagittal images showing complete removal of the fourth ventricular tumor.

then incised. The underlying tumor was immediately evident, as it was adherent to the deep surface of the vermis and roof of the fourth ventricle. The tumor was resected with an aspirator superiorly, inferiorly and then laterally to both sides. The fourth ventricle was widely exposed in its entirety and the brainstem was unharmed.

The patient awoke from surgery with some truncal ataxia but more significantly, densely mute. Because of dependence on the EVD, a ventriculoperitoneal shunt was implanted 2 weeks afterward. The final result from all the pathological analysis was grade I ependymoma, and because the tumor had been fully removed, neither radiation nor chemotherapy was deemed appropriate or necessary (**Fig. 32.19**). Her speech gradually improved. At one year after her transvermian surgery, she was able to talk, but very slowly. At 4 years after surgery, her speech remained dysarthric and slow. Her tumor has not recurred at last follow-up.

■ Conclusion

The "salvage" operation was necessary in this case, because the tumor portion adherent to the fourth ventricular roof was completely concealed despite a well-executed telovelar approach. It is possible that an even wider dissection of the telovelar junction, further out laterally, would have revealed the obscured portion. This case adds further evidence that cerebellar mutism is causally related to the transvermian approach, as the patient's speech was perfect after the first (telovelar) surgery and destroyed by the second (transvermian).

As more evidence like that accumulates, fourth ventricular surgery may evolve to be dominated by the telovelar approach, and the transvermian approach, once the "workhorse" for midline cerebellar surgery, might be reserved and performed in a limited way, for situations, like the one described here, where the telovelar approach was inadequate, and the patient's clinical status required a "salvage" operation.

■ Recommended Reading

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